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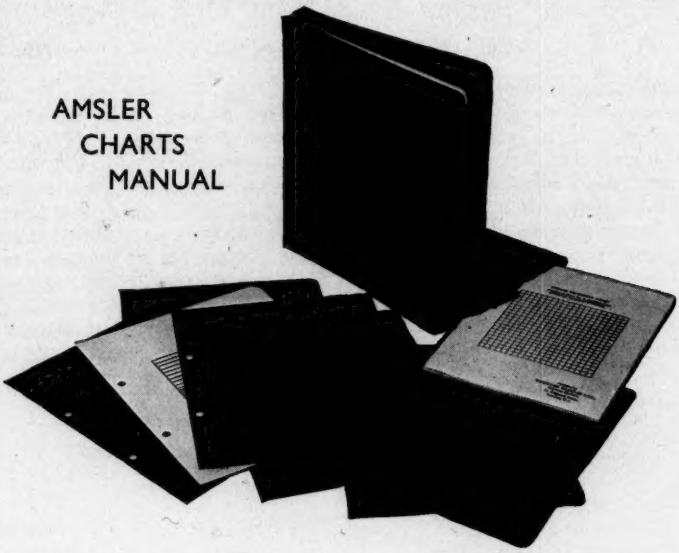

These charts, designed by Professor Marc Amsler of Zurich, make it possible to detect, localise, and analyse the visual functional disturbances which accompany the beginning and evolution of maculopathies. (The word "maculopathy," analogous to "retinopathy," is used by Professor Amsler to express all pathological conditions of the macula.) Many functional disturbances affecting the central and paracentral region of the field escape the usual quantitative test of central vision since it does not really bring into account the quality of the visual trouble. By means of these charts and the method and technique of examination described by Professor Amsler such qualitative measurements are easily made.

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129A

Brit. J. Ophthal. (1957) **41**, 129.

COMMUNICATIONS

PROGNOSIS IN CASES OF STRABISMUS WITH SPECIAL REFERENCE TO ORTHOPTIC TREATMENT*

BY

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WHEN orthoptic treatment was first practised it was thought that, if it were applied in cases of non-paralytic strabismus where there was good visual acuity in each eye, restoration of normal binocular function would invariably result. In the early 1930s it was not unusual for children suffering from squint to be subjected to countless sessions of treatment with reflecting stereoscopes in the hope of establishing fusion. It was not until Chavasse (1939) published his theories with regard to the binocular reflexes and their development, that the true limitations of orthoptic treatment became more widely recognized. The important fact then emerged that, the earlier the onset of a constant strabismus and the longer the interval of time before corrective treatment is carried out, the less likely is it that normal binocular function will be restored, no matter how much orthoptic treatment is given.

Experience with a large series of cases of strabismus and the study of the work of Chavasse have taught us that in certain cases binocular single vision cannot be attained whatever the treatment, while in others, provided the correct treatment is given, the restoration of binocular single vision is certain. Thus, statistics regarding the results of treatment in terms of binocular function are quite meaningless unless they contain information about the types of case under review. The correct treatment varies according to the type of case; it may consist merely of appropriate spectacles (with or without the assistance of orthoptic treatment), or of surgical treatment (with or without the assistance of spectacles and orthoptic treatment), or of orthoptic treatment only. Despite these facts, in many clinics a routine course of orthoptic treatment is still recommended and carried out indiscriminately in all cases of non-paralytic strabismus. The result is that a large percentage of patients treated by orthoptic measures fail to regain normal binocular function. The conclusion is drawn that in these cases orthoptic treatment has failed, as indeed it has, but the failure is due to its employment where it was not indicated.

After studying the history of a case of strabismus and after carrying out a thorough examination, it should be possible in most instances to decide

* Received for publication December 4, 1956.

what sort of treatment is needed and to give a fairly accurate prognosis with regard to the possibility of restoring binocular single vision. Admittedly, in young infants too uncooperative for examination on a major amblyoscope, the prognosis may have to be guarded, but even then, with a knowledge of the case history and the family history and a simple clinical examination, a reasonably accurate prognosis can often be given.

It is our aim in this article to review some of the factors concerned in prognosis and to consider how appropriate treatment can be planned with special reference to the place of orthoptic measures.

(A) NON-PARALYTIC STRABISMUS

I. Non-paralytic Convergent Strabismus*

Perhaps it is chiefly in convergent strabismus that the selection of cases suitable for orthoptic treatment appears to present the most difficulty. It was therefore decided to submit to critical analysis a series of unselected cases of non-paralytic convergent strabismus in children under 10 years of age in order to find out how and where orthoptic treatment appeared to have helped in establishing normal binocular function, and where it had apparently been a waste of time and effort.

An analysis of 287 consecutive cases of non-paralytic convergent strabismus treated at the High Holborn branch of the Moorfields Eye Hospital during the years 1950 to 1954 showed that binocular single vision was restored in 98 cases (34 per cent.) and that 189 cases (66 per cent.) remained non-binocular.

These bare facts, however, do not tell us in which particular type of case a binocular result was obtained and in which type of case there was failure to establish binocular vision. Furthermore, these figures give no hint as to how such factors as the age at onset of squint and the interval between the onset of squint and the treatment that finally corrected it may have affected the prognosis.

If we divide the 287 cases into accommodative and non-accommodative, we see at once how the prognosis differs (Table I).

TABLE I

NON-PARALYTIC CONVERGENT STRABISMUS

Type of Strabismus	Accommodative	Non-Accommodative	Total
Degree of Binocular Single Vision	Fully binocular 61 (82%)	Binocular 24 (11%)	85
	Binocular with intermittent convergence for near fixation 13 (18%)	Non-binocular 189 (89%)	202
Total	74	213	287

* We have used the term non-paralytic rather than concomitant because few cases of convergent strabismus are strictly concomitant, a concomitant strabismus being one in which the angle of deviation remains the same whichever eye is fixing and in whichever direction the eyes are looking.

(1) NON-PARALYTIC ACCOMMODATIVE CONVERGENT STRABISMUS (74 cases: Tables II and IIA)

> "It seems highly probable that if glasses were prescribed for the infant immediately and as a matter of extreme urgency at the first momentary glide of the eye, the problem of accommodational squint, both in its visual and aesthetic aspects, would disappear. Operations would not be necessary. Multitudes of eyes would be saved for acute vision against the possible loss, by accident or disease, of their fellows. A heavy financial burden would thus be lifted from the community."

So wrote Chavasse (1939), and there is no doubt that his words are very largely true.

Accommodative strabismus rarely appears before the development of the frontal accommodation-convergence reflex, and its incidence is highest in children of the 3-year age group. At this age a child begins to take an increasing interest in small objects situated near at hand and, if he is moderately hypermetropic and in consequence has to exercise considerable accommodation in order to see clearly, he may over-converge when he accommodates (since accommodation and convergence act synergically). Although intermittent in the early stages, an accommodative strabismus in which an occasional but marked convergence appears is often more noticeable at the onset than is a strabismus of the non-accommodative type in which a small static deviation may be overlooked for many months or even years. If a full correction of the refractive error is prescribed at the earliest opportunity, the visual axes are usually restored to parallelism, harmony between accommodation and convergence is re-established, and amblyopia and loss of binocular function are rare. In neglected cases, however, correction of the refractive error may merely reduce but not eliminate the deviation and the therapeutic problem is more complex.

The restoration of binocular single vision in the 74 cases of accommodative strabismus under consideration suggests that there is little to criticize in the treatment they received, although the part played by orthoptic treatment in the results achieved may be open to inquiry.

These 74 cases may be sub-divided into three groups (Tables II and IIA, overleaf):

(*a*) *Fully Accommodative Strabismus* (33 cases).—Here there was no deviation for near or for distance when glasses were worn which corrected the hypermetropia, but there was a convergent deviation without glasses.

(*b*) *Accommodative Strabismus with Convergence Excess* (21 cases).—Here there was no deviation for distance when glasses were worn which corrected the hypermetropia, but there was a constant deviation for near, and a constant convergent strabismus without glasses.

(*c*) *Partially Accommodative Strabismus* (20 cases).—Here there was a constant convergent deviation, the angle of which was greater for near fixation. The angle of deviation was diminished with glasses.

There appeared to be little difference in the aetiology of the three types of accommodative strabismus. In the majority of cases the onset was intermittent with no obvious cause. In all three groups the average age at onset of strabismus was just over 3 years, and the average interval between the onset of the strabismus and the correction of the refractive error was not less than about one year (Table II).

TABLE II

ACCOMMODATIVE CONVERGENT STRABISMUS

Type of Strabismus	Fully Accommodative (33)	Convergence Excess (21)	Partially Accommodative (20)
Average age of Patients (yrs)	5·9	6	6
Average Age at Onset of Squint (yrs) ...	3·1	3·6	3·2
Average Interval between Onset of Squint and Correction of Refractive Error (yrs)...	2	1	1

(*a*) *Fully Accommodative Strabismus* (33 cases).—Restoration of full binocular single vision was effected in ten of the 33 cases by correction of the refractive error alone without the aid of orthoptic treatment. Of the remaining 23 cases in which orthoptic treatment was given, there was no appreciable loss of binocular function (with the exception of three cases in which amblyopia had first to be rectified by occlusion of the non-squinting eye). If orthoptic treatment had been omitted, there is no reason to suppose that the ultimate result would have been less successful provided adequate attention was given to the prescription and wearing of glasses.

It is generally agreed that a child with a fully accommodative strabismus should be taught to relax his accommodation when not wearing glasses, thus establishing binocular single vision at the expense of clear vision. Much time is often spent in teaching such a child relative accommodation (*i.e.* clearing the image whilst maintaining single vision). In a child with a refractive error of about +3 D sph. or less, whose refractive error is likely to decrease with age, binocular single vision without glasses will no doubt be achieved without the aid of the synoptophore, bar-reader, and other orthoptic apparatus *provided the strength of the glasses is gradually reduced as and when indicated.*

In a child with hypermetropia of more than about +3 D sph. or with considerable astigmatism, who is likely to continue wearing glasses indefinitely, it is questionable whether the time that is often spent in teaching him to bar-read and to improve binocular visual acuity without glasses (or with a reduced correction) is really justifiable. It is usually preferable to give the patient a few lessons on how to relax accommodation and convergence

without glasses so as to enable him to maintain a reasonable ocular appearance when he is unable or does not wish to wear glasses.

(*b*) *Accommodative Strabismus with Convergence Excess* (21 cases).—In the convergence excess and partial accommodative type of strabismus, the child may continue to over-converge for near vision even after prolonged orthoptic treatment. Although in the passage of time we have found that in some cases this over-convergence may tend to disappear, this is the exception rather than the rule.

Opinions differ as to the treatment of children with accommodative strabismus of this type, and there are three current methods:

(*i*) Full correction of the refractive error combined with orthoptic treatment. Supporters of this method believe that in time the relationship between accommodation and convergence will right itself.

(*ii*) Bifocal spectacles. This method is rarely used at this hospital and we have therefore scant experience of its value, but we have often observed that children who are wearing bifocals frequently fail to accept the extra "plus correction" and tend to read through the distance part of the glasses, thus defeating the purpose of the bifocal glasses. It is of course an obvious fact that bifocal glasses are useless unless the extra "near-vision" addition does actually secure binocular fixation.

(*iii*) Surgery, usually after a trial period of orthoptic treatment. This in our experience has been the most satisfactory method. It has been suggested that operation may cause consecutive divergence in later life, but this is unlikely provided binocular functions are strong.

In this series of cases, patients who had orthoptic treatment, particularly when combined with operation, show better results than those who did not. Although there is no doubt that in these cases orthoptic treatment played a useful part in re-establishing a normal relationship between accommodation and convergence, it is not possible to lay down hard and fast rules as to its use and each case should be judged on its merits. Insufficient time has elapsed to consider the possibility of consecutive divergence, but early results show no tendency to divergence. Indeed one would not expect divergence to occur in view of the good binocular function present. Re-examination of patients in a few years time may give valuable information as to the advisability of surgery in this type of case.

(*c*) *Partially Accommodative Strabismus* (20 cases).—The importance of dealing immediately with the refractive error is shown in the cases of partially accommodative strabismus in which optical treatment has failed to eliminate the deviation entirely or has been inadequately carried out, for it is probable that many of the cases in this group were originally cases of fully accommodative strabismus or of accommodative strabismus with convergence excess. Although the deterioration of binocular function which occurred in ten cases was not as severe as is found in neglected cases of non-accommodative

convergent strabismus, it was sufficient to complicate and lengthen the treatment.

Operation is invariably necessary in this type of strabismus. Although full binocular single vision was restored in five cases by surgery alone, orthoptic treatment was undoubtedly of value in consolidating the results of the operation in fourteen cases.

In one case binocular single vision was restored by orthoptic treatment alone.

Table IIA summarizes the results of treatment in all three groups:

TABLE IIA

RESULTS IN 74 CASES OF ACCOMMODATIVE CONVERGENT STRABISMUS

Treatment	Restoration of Full Binocular Single Vision			Restoration of Binocular Single Vision with Intermittent Convergence for Near		Total
	Fully Accommodative	Convergence Excess	Partially Accommodative	Convergence Excess	Partially Accommodative	
Operation Only ...	—	1	5	3	1	10
Operation and Orthoptic Treatment ...	—	8	10	1	3	22
Orthoptic Treatment and Glasses ...	23	4	—		1	31
Glasses Only ...	10	—	—	1	—	11
	33	13	15	8	5	
Total	61 (82%)			13 (18%)		74

(2) NON-PARALYTIC NON-ACCOMMODATIVE CONVERGENT STRABISMUS (213 cases: Table III, opposite)

In the non-accommodative group 165 patients had orthoptic treatment, 138 with operation, and 27 without operation. In all cases the visual axes had been made parallel (or approximately so) by operation, except where the deviation was negligible or was corrected by glasses.

Of the total 213 cases, 24 (11 per cent.) regained binocular single vision; in these a favourable result had been envisaged because the average age at onset of squint was 4·8 years.

The remaining 189 cases (89 per cent.) were classified as non-binocular. In 116 (61 per cent.), there was no improvement of binocular function and orthoptic treatment proved to be of no value. Had an early and accurate prognosis been made, much time and effort could have been saved in their treatment. (It will be observed that seventy of the 116 patients had orthoptic treatment and an operation, and that fifteen of them had orthoptic treatment only.) In 73 cases (39 per cent.) there was some improvement in binocular function (*i.e.* normal retinal correspondence was established with doubtful

TABLE III

NON-PARALYTIC NON-ACCOMMODATIVE CONVERGENT STRABISMUS

Treatment	Restoration of Binocular Single Vision	Non-Binocular		Total
		Subnormal Binocular Vision	Defective Binocular Function*	
Operation Only	8	9	31	48
Operation and Orthoptic Treatment	16	52	70	138 } 165
Orthoptic Treatment Only	—	12	15	27 }
		73 (39%)	116 (61%)	
Total	24 (11%)	189 (89%)		213
Average Age of Patients (yrs)	6·7	6·5	5·5	
Average Age at Onset of Squint (yrs)	4·8	2·2	1·4	
Average Time between Onset of Squint and Corrective Treatment (yrs)	1·9	4	3·5	

* Severe amblyopia 25, lack of normal retinal correspondence 73, abnormal retinal correspondence 16, nystagmus 2, Total 116.

fusion), and it may be concluded that orthoptic treatment may have been of value. These cases have been classified as having "subnormal binocular vision"; they cannot be considered as cured nor should they be regarded as complete failures, for a subnormal form of binocular vision has been established which may result in improvement in visual judgement and may prevent consecutive divergence. Furthermore, it is known that in this type of case binocular single vision may develop spontaneously after a few months or years. The routine re-examination of a series of such cases after an interval of a few years may show that spontaneous recovery is more common than is supposed. The following is an example of such a case:

Case 1, a boy aged 9 years, had a left convergent strabismus which was first noticed at the age of about 5 years. He had had two operations (at the age of 7 years) and orthoptic treatment before attending this hospital.

Examination

Visual Acuity: Right 6/9; with +2·25 D sph., +1 D cyl., axis 90°, 6/6 and N5.
Left 6/18; with +3 D sph., +1·75 D cyl., axis 90°, 6/9 and N5.

Cover Test: Constant left convergent deviation with and without glasses for both near and distance.

Major Amblyoscope: Angle with glasses +11°, R/L 2△
Angle without glasses +13°, R/L 1△
Doubtful fusion.

Treatment: Right medial rectus recession at the age of 11 years.

Post-operative Examination

Visual Acuity: With glasses Right 6/6 and N5; Left 6/6 and N5.

Major Amblyoscope: Angle with glasses +2°, R/L 1△
Angle without glasses +12°, R/L 1△
Doubtful fusion.

Examination 3 years later at the age of 15

Cover Test: Latent convergence for near and distance with glasses.
Constant left convergent deviation without glasses.

Major Amblyoscope: Angle with glasses +2° ⊖
Angle without glasses +10° ⊖
Fusion and stereoscopic vision present.

In many cases one can reach a decision regarding the prognosis upon the information obtained from the case history (assuming this to be correct). Thus, in the above case, the prognosis was good because the squint did not occur until the age of 5 years (at which age the binocular reflexes are fairly well developed).

The importance of taking a full history cannot be over-emphasized, for it may provide information as to the type of strabismus present, the treatment required, and the likely result of such treatment. Inquiry having been made as to direction of the deviation, whether unilateral or alternating, and whether constant or intermittent, it is important to ascertain:

(*a*) Age at onset of strabismus,
(*b*) Mode of onset,
(*c*) Heredity.

(*a*) *Age at Onset of Strabismus and Duration of Strabismus.*—According to Chavasse, the unconditioned binocular reflexes are in a state of flux during the first five years of life, not becoming fully grounded until about the eighth year. Constant interruption of binocular vision by a sensory or motor obstacle will therefore almost invariably result in permanent arrest of their development unless corrective treatment (*i.e.* the alignment of the visual axes by surgery or by other means) is undertaken at an age when the re-establishment and stabilization of normal binocular function is still possible.

The significance of the age at onset of strabismus is shown in the following case:

Case 2, a girl aged 8 years, had had a mild pyrexia and a "lump" at the side of her neck 5 weeks previously when a sudden and constant right convergent strabismus was noticed.

Examination

Visual Acuity: Right 6/5 and N5; Left 6/5 and N5. No refractive error.
Cover Test: Constant right convergent strabismus.
Ocular Movements: Normal. No relevant refractive error.
Major Amblyoscope: Angle +18° R/L 5△ fixing with either eye.
Fusion range +18° to +6°, stereopsis present.

Treatment: after 5 months the condition was unchanged and a right medial rectus recession and a right lateral rectus resection were carried out.

Post-operative Examination

Cover Test: Latent convergence for near with rapid recovery.
No deviation for distance.

Major Amblyoscope: Angle +5° L/R 1△
Fusion range +15° to −5°, stereopsis present.

The binocular reflexes had become fully grounded before the onset of the strabismus at the age of 8 years. Orthoptic treatment was unnecessary post-operatively as insufficient time had elapsed between the onset of the strabismus and its correction by surgery for deterioration in binocular function to occur.

The following case illustrates how immediate treatment in early childhood may result in a functional cure:

Case 3, a girl aged 4 years, had had a gradual onset of left convergent strabismus noticed at the age of 3 years. At first the deviation was intermittent.

Examination

Visual Acuity: Right 6/18; with +1·25 D sph., 6/12 and N5.
Left 6/18; with +1·25 D sph., 6/12 and N5.

Cover Test: Constant alternating convergent strabismus with and without glasses.

Major Amblyoscope: Angle with glasses +25/30° ⊖
Angle without glasses +30° ⊖
Patient too young for assessment of state of binocular vision.

Treatment: (1) Left medial rectus recession, left lateral rectus resection, and (2) right medial rectus recession at the age of 4 years. She attended for twelve sessions of orthoptic treatment post-operatively.

Re-examination: at the age of 6 years the clinical findings were as follows:

Visual Acuity: Right 6/5 and N5; Left 6/6 and N5. No glasses worn.
Cover Test: No deviation for near and distance.
Maddox Rod: Eso 2△ ⊖ fixing with either eye.
Maddox Wing: Exo 2△ ⊖
Major Amblyoscope: Angle 0° ⊖ fixing with either eye.
Fusion range +8° to −4°, stereopsis present.

In this case the following factors led one to anticipate a good result:

(*i*) Onset of strabismus comparatively late, *i.e.* at a time when the binocular reflexes had already undergone normal early development.
(*ii*) Short duration of the strabismus before the corrective treatment was undertaken which restored parallelism of the visual axes.
(*iii*) Deviation intermittent at first.

Orthoptic treatment was limited to twelve sessions, sufficient to consolidate the effects of surgery.

The following example shows how loss of binocular vision can become irreparable if the strabismus is of early onset and if corrective treatment is delayed:

Case 4, a girl aged 3½ years, had had alternating convergent strabismus since birth.

Examination

Visual Acuity: Right 6/24; with +2·50 D sph., +0·50 D cyl., axis 90°, 6/12 partly.
Left 6/18; with +2·50 D sph., 6/12.

Cover Test: Constant alternating convergent strabismus with and without glasses, but prefers to fix with the left eye.

Major Amblyoscope: Angle with glasses +25° ⊖
Angle without glasses +32° ⊖
Patient too young for assessment of state of binocular vision.

Treatment: Right medial rectus recession, and right lateral rectus resection at the age of 3½ years. She attended for twelve sessions of post-operative orthoptic treatment.

Re-examination: At the age of 8 years the clinical findings were as follows:

Visual Acuity: Right 6/9 and N5; Left 6/9 and N5. No glasses worn.
Cover Test: Constant right convergent strabismus.
Major Amblyoscope: Angle +8° ⊖ fixing with either eye.
Lack of normal retinal correspondence.

Although no time was lost in correcting the strabismus by operation after the child was first brought for examination, the onset of the strabismus at birth and the loss of the vital early years of binocular development had led to a complete loss of binocular vision. The post-operative orthoptic treatment was presumably given in the hope of effecting a functional cure, but in view of the history it was virtually a waste of time.

On the other hand, there are cases in which a spontaneous recovery with binocular single vision occurs post-operatively in spite of a somewhat adverse history and pre-operative findings.

Case 5, a boy aged 7 years, had had alternating convergent strabismus since the age of 3 or 4 months. The strabismus was intermittent at first but had become constant by the age of 2½ years. Glasses prescribed at the age of 4 years did not improve the squint.

Examination

Visual Acuity: Right 6/18; with −0·5 D sph., +1·5 D cyl., axis 90°, 6/12.
Left 6/12; with −0·5 D sph., +1 D cyl., axis 90°, 6/9.
Cover Test: Constant alternating convergent strabismus with and without glasses.
Major Amblyoscope: Angle with glasses fixing right +42° R/L 1△
Angle with glasses fixing left +40° R/L 6△
Lack of normal retinal correspondence.

Treatment: (1) Right medial rectus recession, right lateral rectus resection, and (2) left medial rectus recession at the age of 8 years.

Post-operative Examination

Visual Acuity: Right 6/9 and N5; Left 6/9 and N5. With glasses
Cover Test: Constant alternating convergent strabismus with and without glasses.
Major Amblyoscope: Angle with and without glasses fixing right + 12° ⊖
fixing left + 10° ⊖
Normal retinal correspondence was present but no fusion.

Re-examination: 3 years later he had binocular single vision and his condition was as follows:

Cover Test: Latent convergence for near and distance with moderate recovery.
Maddox Rod: Fixing right Eso 7△ ⊖; fixing left Eso 8△ ⊖
Maddox Wing: Eso 5/7△ ⊖
Major Amblyoscope: Angle +3° ⊖ fixing with either eye.
Fusion range +22° to 0°, stereopsis present.

This case is not exceptional. The history was important. The fact that the squint was intermittent at first and did not become constant until the age of 2½

years was strong evidence that binocular function had been basically good in spite of the lack of normal retinal correspondence and therefore with accurate surgical treatment the prognosis was good.

(*b*) *Mode of Onset.*—It is interesting to note that in the 74 cases of accommodative strabismus in which the onset was slow and intermittent, there was a 100 per cent. restoration of binocular single vision. This success is probably also due to the comparatively late average age of onset of the strabismus and the relative ease and rapidity with which the visual axes can be restored to parallelism in most cases (usually by correction of the refractive error).

In many of the non-accommodative cases, the history of the mode of onset was indefinite, and its influence on the final result was therefore inconclusive.

(*c*) *Heredity.*—A family history of strabismus is not uncommon in accommodative strabismus and in other types related to the state of refraction. It is also found in the non-accommodative type of convergent strabismus in which there may be an hereditary weakness of one or more of the extrinsic ocular muscles.

Our numbers are too small for conclusive evidence, but a family history of squint was found in 39 per cent. of the accommodative group, and in 41 per cent. of the non-accommodative group. It is interesting to note that an hereditary factor was found more often in the "non-binocular" cases than in those in which binocular single vision was regained.

Defective Binocular Function (116 cases).—Table III shows that 116 patients were discharged with defective binocular function (severe amblyopia, 25; lack of normal retinal correspondence, 73; abnormal retinal correspondence, 16; and latent nystagmus, 2). 85 of these patients had had orthoptic treatment, in seventy cases combined with operation. An analysis of these cases indicates why there was no response to orthoptic treatment:

(*a*) *Amblyopia.*—In 54 cases (25 per cent.) of the non-accommodative group, the visual acuity of the squinting eye was less than 6/36 at the first examination. Total occlusion of the fixing eye was carried out in 48 cases, but the remaining six were considered unsuitable for occlusion owing to the relatively mature age of the patients and the early onset of the squint.

After an adequate period of occlusion the 48 cases were classified as follows according to the results obtained:

Group 1.—Twenty cases (41 per cent.), in which equal or nearly equal visual acuity was obtained in each eye.

Group 2.—Nine cases (19 per cent.), in which the visual acuity of the squinting eye improved only to 6/18.

Group 3.—Nineteen cases (40 per cent.), in which there was no improvement in the visual acuity of the squinting eye.

There was no great difference between the average age at onset of the strabismus and the average interval between onset and the commencement of occlusion, but the average age of the patients in the first group was

significantly lower than that of those in the second and third groups. The results of occlusion in these 48 cases are shown in Table IV.

TABLE IV
RESULTS OF OCCLUSION IN 48 CASES OF AMBLYOPIA

Visual Acuity after Occlusion	Average Age of Patient (yrs)	Average Age at Onset of Strabismus (yrs)	Average Time between Onset of Strabismus and Occlusion (yrs)
1. Equal or nearly equal ...	4·9	1·6	3·3
2. Squinting eye did not improve beyond 6/18	5·5	2·1	3·0
3. Squinting eye did not improve	5·7	1·5	3·1

Thus amblyopia which did not respond adequately to occlusion certainly accounted for 25 cases and probably also for an additional nine cases in which there was failure to restore binocular single vision. The desirability of carrying out occlusion as early as possible is emphasized by this analysis.

We have not yet had any experience of the treatment of amblyopia by the after-image method of Cüppers. This method appears to offer great possibilities for the improvement of vision in cases of suppression amblyopia, especially those associated with eccentric fixation.

(*b*) *Lack of Normal Retinal Correspondence.*—73 patients showed lack of normal retinal correspondence; 45 of them had orthoptic treatment (combined with operation in 41 cases) and in no case was there any improvement in binocular function. 28 patients had surgical treatment without any orthoptic treatment. The age and time factors are much the same both in those patients who had orthoptic treatment and those who did not (Table V).

TABLE V
PATIENTS WITH LACK OF NORMAL RETINAL CORRESPONDENCE

Treatment	Operation Only	Orthoptic Treatment (+Operation in 41 Cases)	Total
No. of Patients	28	45	73
Average Age of Patients at Operation (yrs)...	5	5·7	
Average Age at Onset of Squint (yrs)	1	1·7	
Average Time between Onset of Squint and Corrective Treatment (yrs)	3·7	3·7	

The average age at onset of squint in the patients in this group is notably early. It seems quite clear that in these cases pre-operative orthoptic treatment is useless and it is also doubtful whether post-operative orthoptic treatment is of any value.

(*c*) *Abnormal Retinal Correspondence.*—This was present in only sixteen (8 per cent.) of the 213 cases of non-accommodative convergent strabismus.

The differentiation between a definite abnormal retinal correspondence and a lack of normal retinal correspondence with a tendency towards anomalous correspondence requires careful clinical investigation. In the former condition, when tested on a major amblyoscope, the patient "puts the lion into the cage" at an angle less than the objective angle of deviation; a small range of fusion on non-corresponding points of the retinae is usually present, and the response to the after-image test is consistently abnormal. In the latter condition, the patient attempts to "put the lion into the cage" at an angle differing from the true angle of deviation (as measured objectively), but is unable to do so on account of suppression; there is no range of fusion and the response to the after-image test may be normal or abnormal.

In this series, only cases with a definite angle of anomaly and a small range of fusion have been classified as showing abnormal retinal correspondence.

It is not possible to draw any definite conclusions from so small a number, but of the sixteen cases of abnormal retinal correspondence in which orthoptic treatment was carried out (ten combined with operation) not one developed normal retinal correspondence. The time spent in striving to achieve normal retinal correspondence seems to be fruitless, especially since it is now generally believed that true abnormal retinal correspondence is a perverted form of binocular single vision and has a relative stability. Post-operative consecutive divergence is less likely to occur in this condition than in cases in which the perversion has not become strongly established. It should be mentioned, however, that in the past abnormal retinal correspondence was much more loosely defined, and that cases alleged to have been cured by surgical and orthoptic treatment were probably examples of lack of normal retinal correspondence.

The essential age and time factors in the history of the sixteen cases of abnormal retinal correspondence are somewhat similar to those of the 73 patients with lack of normal retinal correspondence (Table VI).

TABLE VI
PATIENTS WITH ABNORMAL RETINAL CORRESPONDENCE

Treatment	Operation Only	Orthoptic Treatment (+ Operation in 10 Cases)	Total
No. of Patients	1	15	16
Average Age of Patients (yrs) ...	11	6·7	
Average Age at Onset of Strabismus (yrs)...	2	1·4	
Average Time between Onset of Strabismus and Corrective Treatment (yrs)	8	3·5	

(*d*) *Latent Nystagmus.*—Cases in which gross latent nystagmus is associated with convergent strabismus need no special comment. Defective binocular function or binocular instability is invariably present.

Subnormal Binocular Vision (*73 cases*).—By studying the history of the case, taking the age of the patient into consideration, and making a few simple tests, it should not be difficult to differentiate between patients who are likely to regain full binocular single vision, either by operation alone or with the aid of orthoptic treatment and occlusion if necessary, and those who are unlikely to attain binocular vision.

There is however an intermediate group which consists of those who improve to the extent of regaining equal or nearly equal visual acuity, normal retinal correspondence, and doubtful fusion. These patients may benefit from the improved visual judgment which they thereby gain, and, providing the angle of deviation is small, they may develop binocular single vision spontaneously with the passage of time. This group may indeed be considerably helped by orthoptic treatment; 73 of the 213 cases of non-accommodative convergent strabismus belonged to this group.

In comparing these with the 116 cases with defective binocular function, Table III shows that, although, as one would expect, the average age at onset of strabismus was lower in the patients with defective binocular function, the average interval between the onset of strabismus and the commencement of corrective treatment was slightly shorter.

It is perhaps significant that, of the 73 cases with subnormal binocular vision, 21 (30 per cent.) already had normal retinal correspondence before treatment started, and five (7 per cent.) were too young for an accurate assessment of the state of binocular vision. At the first examination the remaining 47 patients (63 per cent.) had no binocular vision, either through gross amblyopia or lack of normal retinal correspondence. (There were no cases of abnormal retinal correspondence in this group.)

The average age at onset of strabismus was 2·2 years in patients with subnormal binocular vision, compared with 1·4 years in the patients with defective binocular function who did not respond to treatment. The average interval between the onset of the strabismus and the commencement of corrective treatment was 4 years and 3·5 years respectively, and the average age of the patient when treatment was begun was 6·5 and 5·5 years respectively (*Table III*).

Since there appears to be no outstanding difference in the general trend of the history of the two groups, it may be that in some cases, although the loss of normal binocular vision seems apparent pre-operatively, surgical correction of the strabismus may induce a return of weak binocular vision which had already existed before the onset of the strabismus.

In seven cases normal retinal correspondence developed spontaneously after operation.

Vertical Factor in Convergent Strabismus.—In fifty (24 per cent.) of the 213 *non-accommodative* cases there was a primary vertical deviation due to defective action of one or more of the vertically acting muscles. Unlike certain cases of convergent strabismus in which there is over-elevation of the non-fixing eye on adduction which becomes rectified when the horizontal deviation is corrected, in these cases surgical correction of the horizontal deviation fails to reduce the vertical deviation. Thus, if binocular single vision is to be expected, operation on the vertically acting muscles becomes necessary.

The history of these cases appears to be similar to that of the cases with a purely horizontal deviation and the results as regards binocular function are also similar.

Follow-up Investigations.—Of the 213 cases of non-paralytic non-accommodative convergent strabismus under investigation, ninety were re-examined 3 years after discharge from the orthoptic department in order to find out if there was:

(*i*) Any deterioration of binocular function.
(*ii*) Any improvement of binocular function.
(*iii*) Any evidence of consecutive divergence.

These cases were chosen at random and comprised the following:

6 discharged with binocular single vision.
34 discharged with subnormal binocular vision.
50 discharged with defective binocular function.

The essential facts are set out in Table VII:

TABLE VII
CONDITION OF PATIENTS

On Discharge			On Re-examination	
Full Binocular Single Vision		6	Full Binocular Single Vision	6
Subnormal Binocular Vision	Fusion with no Amplitude	11	Binocular Single Vision for Distance with good Fusion Amplitude	1
			Fusion with no Amplitude	10
	Simultaneous Macular Perception	23	Fusion with no Amplitude	3
			Simultaneous Macular Perception	18
			Lack of Normal Retinal Correspondence (one divergent)	2
	Total	34	Total	34
Defective Binocular Function	Abnormal Retinal Correspondence	8	Abnormal Retinal Correspondence	5
			Lack of Normal Retinal Correspondence (one divergent)	2
			Fixation Disparity	1
	Lack of Normal Retinal Correspondence	33	Lack of Normal Retinal Correspondence (one divergent)	29
			Simultaneous Macular Perception	4
	Amblyopia	9	Amblyopia (one divergent)	9
	Total	50	Total	50

Of the 34 cases discharged with subnormal binocular vision, there was no change in binocular function in 28, but consecutive divergence was present in one case. In four cases there had been an improvement in binocular function to the extent of restoration of binocular single vision for distance in one case. There was however, little evidence in the history of these cases to suggest the likelihood of an improvement of binocular function in the passage of time except in one case in which the age of onset of the strabismus was said to be 5 years.

There was no improvement in binocular function in 45 of the fifty cases with defective binocular vision. Consecutive divergence was present in three cases. Four patients who originally had lack of normal retinal correspondence had developed normal retinal correspondence, and one patient with abnormal retinal correspondence had developed fixation disparity. Here again there was no indication in the history leading one to anticipate an improvement in binocular function. It is not, however, possible to draw conclusions as to the probability of further spontaneous improvement of binocular function in so few cases.

We have seen that, in the non-accommodative type of convergent strabismus, the chances of restoring full binocular single vision are remote unless the strabismus is of later onset (*i.e.* after about the 2nd year when the binocular reflexes are fully or partially developed) and is speedily dealt with before deterioration of binocular function can take place. The importance of orthoptic treatment in the re-education of normal binocular function should not be under-estimated in cases in which the history and clinical findings show that this is potentially possible, and careful consideration before undertaking treatment should lead to considerable lessening of unnecessary work.

II. Primary Divergent Strabismus

In dealing with divergent strabismus we have included adult patients, since especially in intermittent divergence, the patient may not suffer from symptoms until adult life. Early treatment in childhood is however to be recommended.

A consecutive series of 121 cases of divergent strabismus has been studied. The cases were divided into two main groups:

(*1*) *Constant Divergent Strabismus.*—This may date from birth or earliest infancy. Binocular vision is frequently lost.

(*2*) *Intermittent Divergent Strabismus.*—The intermittent nature of the squint ensures that normal binocular function is preserved. In neglected cases the deviation may in time become constant with deterioration of binocular function.

(1) Constant Divergent Strabismus (29 cases)

The average age of the patients in this group was 29 years, the average

age at onset of strabismus was 2 years, and the average interval between the onset of squint and corrective treatment was 23·7 years. Weak fusion was demonstrable in three cases pre-operatively. In the remaining 26 cases, gross amblyopia was present in nine and complete loss of binocular vision in seventeen.

In nine cases binocular single vision was restored post-operatively. In three of these fusion could not be demonstrated immediately after operation, but the deviation appeared to be latent with an apparent recovery to binocular fixation as shown by the cover test. Of the other six cases in which binocular single vision developed, two showed weak fusion pre-operatively and four showed complete loss of binocular vision. In the two cases discharged with an intermittent deviation there was little amplitude of fusion and no stereopsis. In a follow-up examination 3 years later, re-divergence had occurred in one of these cases with deterioration of binocular function.

Post-operative spontaneous development of normal binocular function in cases in which there is no pre-operative evidence of normal retinal correspondence is not uncommon in divergent strabismus as the following case exemplifies:

Case 6, a woman aged 29, had had left divergent strabismus probably since birth, which became more noticeable after fits during teething.

Examination

Visual Acuity: Right 6/36; with −2·5 D sph., −2·25 D cyl., axis 100°, 6/5 and N5.
Left 6/60; with +1 D sph., −4 D cyl., axis 50°, 6/24 and N8.
Cover Test: Constant left divergent strabismus.
Major Amblyoscope: Angle fixing right −34° R/L 2△
Angle fixing left −33° L/R 2△
No subjective angle obtained.

Treatment: (1) Left lateral rectus tenotomy and left medial rectus resection.
(2) Right medial rectus marginal myotomy (on account of slight over-correction) and right inferior rectus marginal myotomy.

Post-operatively the deviation was latent and, although simultaneous macular perception was present, fusion was still not demonstrable on the major amblyoscope.

A routine examination 2 years later revealed that since the operation binocular single vision with a good range of fusion and stereopsis had developed and the vision of the left eye had improved from 6/24 to 6/9:

Re-examination

Visual Acuity: Right 6/5 and N5; Left 6/9 and N5. With glasses.
Cover Test: Moderate latent divergence for near and distance with rapid recovery.
Major Amblyoscope: Angle fixing right −4° R/L 3△
Angle fixing left −6° R/L 1△
Fusion range +8° to −6°, stereopsis present.
Maddox Rod: Fixing right Exo 7△ R/L 2△
Fixing left Exo 9△ R/L 2△
Maddox Wing: Exo 14△

(2) INTERMITTENT DIVERGENT STRABISMUS (92 cases)

These cases have been divided into the following groups:

(*a*) *Divergence Excess* (43 cases: Maddox rod reading exceeds Maddox wing reading).

(*b*) *Convergence Weakness* (24 cases: Maddox wing reading exceeds Maddox rod reading).

(*c*) *Mixed* (25 cases: Maddox rod and wing reading approximately the same). These cases are probably originally of the divergence excess type with secondary convergence weakness.

Binocular single vision is maintained either for near (divergence excess type) or for distant vision (convergence weakness type), although in neglected cases the deviation may become constant. The age at onset of the strabismus is often indefinite, varying from birth to adolescence, and in this series averages 4·5 years. The interval between the onset of strabismus and the beginning of corrective treatment is correspondingly indefinite and averages 10·9 years. In general, intermittent divergent strabismus of the divergence excess type starts at an earlier age than that of the convergence weakness type.

Diplopia and other symptoms resulting from the voluntary effort to maintain binocular single vision are common, but in some cases suppression may be intense, making the patient unaware of his squint. The intermittent nature of the strabismus renders it amenable to orthoptic treatment although surgical treatment is invariably necessary to effect a complete cure, with the possible exception of certain cases of the convergence weakness type in which there is sometimes relief of symptoms from orthoptic treatment alone. Controversy still exists over the age at which operation should be performed. Some people disapprove of surgical treatment in childhood believing that the normal development of the face during the years of growth will cause re-divergence later. In this series, 21 children were operated on before the age of 10 years; in fourteen of these full binocular single vision was restored, four patients still had some intermittent deviation at times, and three who had weak or doubtful fusion pre-operatively were discharged with constant convergence and no fusion. During a follow-up examination 3 years later, seven of the fourteen patients who were discharged with binocular single vision and no deviation were re-examined, and their condition was found to be exactly the same. One patient discharged with a constant convergent deviation and no fusion was showing a tendency to re-divergence.

The prognosis in primary divergent strabismus therefore depends mainly on the type of onset, *i.e.* whether it is intermittent or constant. In almost all cases in which the deviation is originally intermittent, binocular single vision can be restored, despite the fact that through neglect the divergence may have subsequently become constant with deterioration of binocular function. Orthoptic treatment is often of value in helping the restoration of binocular function.

If a divergent strabismus is constant from the beginning, especially if the condition started within the first 2 years of life, the prognosis is usually poor, although in a few cases binocular single vision may be restored spontaneously after operation, provided the visual axes are restored to parallelism.

(B) CONGENITAL PARALYTIC STRABISMUS

A patient with a congenital paralytic strabismus may be able to preserve a full or partial field of binocular single vision by adopting a compensatory head posture and/or by possessing an abnormally large vertical fusion range. Thus the condition may sometimes remain compensated throughout life On the other hand symptoms of decompensation may occur in adult life, and if untreated an intermittent squint may gradually give place to a constant deviation. In some cases of paralytic strabismus of severe degree there may be a complete loss of binocular vision from earliest infancy. With the exception of the latter cases, the prognosis in congenital paralytic strabismus is invariably good and depends upon surgical rather than orthoptic treatment, but careful orthoptic investigation provides useful information as to the surgical treatment required.

Our series of 107 patients are divided into two groups:

I. Patients with no binocular vision.
II. Patients whose binocular vision has been maintained.

I. Patients with No Binocular Vision (15 cases)

These fifteen patients have been classified as follows:

(1) Palsy of Horizontally-Acting Muscles (3).

(*a*) Duane's retraction syndrome (2).
(*b*) Moebius' syndrome (1).

(2) Palsy of Vertically-Acting Muscles with Secondary Convergence (7).

(*a*) Convergent strabismus with palsy of an elevator or depressor muscle of one eye.
(*b*) Convergent strabismus with palsy of both elevators of one eye (1).
(*c*) Convergent strabismus with mixed or multiple palsy (2).

(3) Palsy of Vertically-Acting Muscles with Secondary Divergence (5).

(*a*) Divergent strabismus with palsy of both depressor muscles of one eye (1).
(*b*) Divergent strabismus with mixed or multiple palsy (4).

Treatment consisted only of operation to improve the cosmetic appearance. Orthoptic treatment was not given in any case.

II. Patients whose Binocular Vision has been Maintained (92 cases)

Binocular single vision was maintained by the adoption of a compensatory head posture in fifty (54 per cent.), and in the remaining 42 (46 per cent.) the deviation was partly controlled and partial or full binocular single vision

was obtained, the head posture being normal. Subjective symptoms occurred in 35 (73 per cent.) of the patients with a compensatory head posture, and in 38 (86 per cent.) of the patients without a compensatory head posture. The average age of the patients with symptoms was 33 years, and of those without symptoms was 14 years.

(1) PALSY OF HORIZONTALLY-ACTING MUSCLES (16 cases: Table VIII).—The following muscles were involved:

Lateral rectus (9).
Lateral rectus associated with Duane's retraction syndrome (6).
Medial rectus (1).

TABLE VIII
PALSY OF HORIZONTALLY-ACTING MUSCLES IN SIXTEEN CASES

Subjective Symptoms	Present ... ... 8 (5 with compensatory head posture)	Absent ... ... 8 (4 with compensatory head posture)
Post-Operative Result	Binocular Single Vision Symptom-free ... ... 8 (head posture normal)	Binocular Single Vision ... ... 6 (3 originally with compensatory head posture now normal) Still some Intermittent Deviation 2 (1 with compensatory head posture unchanged)
	Total 8	Total 8

Eight adult patients complained of symptoms pre-operatively. All were symptom-free after operation.

Of the eight patients (six adults and two children aged about 7) who had no subjective symptoms, six were discharged as satisfactory post-operatively. Three of these (the two children and one adult) originally had a compensatory head posture which disappeared after surgical correction of the paresis. Two adult patients improved but still showed an occasional deviation. One of these had a compensatory head posture which remained unchanged.

Orthoptic treatment was not considered necessary in any of these cases.

(2) PALSY OF VERTICALLY-ACTING MUSCLES (76 cases)

(*a*) *Palsy of Vertically-Acting Muscles unassociated with a Horizontal Deviation* (63 cases: Table IX): The following muscles were involved:

Superior rectus	31
Superior oblique	25
Inferior oblique	5
Inferior rectus	2

55 patients complained of symptoms pre-operatively. 51 of these were completely relieved of their symptoms after surgical correction of the palsy and in the remaining four cases symptoms were much improved.

Of the eight patients who had no subjective symptoms, seven were discharged with binocular single vision. One child still had some intermittent deviation and may require further operation.

A compensatory head posture was present in thirty cases and became normal in 29 cases (Table IX). In four cases binocular function had deteriorated despite the presence of a compensatory head posture, but after operation full binocular function was restored spontaneously.

TABLE IX

PALSY OF VERTICALLY-ACTING MUSCLES IN 63 CASES

(Unassociated with a Horizontal Deviation)

Subjective Symptoms	Present ... ... 55 (23 with compensatory head posture)	Absent ... ... 8 (7 with compensatory head posture)
Post-Operative Result	Binocular Single Vision Symptom-free 51	Binocular Single Vision ... ... 7 (one with compensatory head posture unchanged)
	Symptoms Improved 4	Still some Intermittent Deviation 1
	Total 55	Total 8

(*b*) *Palsy of Vertically-Acting Muscles with Secondary Divergence* (9 cases).—The following muscles were involved:

Superior rectus	7
Superior oblique	1
Inferior rectus	1

Eight patients complained of symptoms pre-operatively. Six were symptom-free after operation and symptoms were considerably improved in the remaining two cases. Five of these eight patients had a compensatory head posture before operation which disappeared after operation in three cases.

One child who had no symptoms but had a compensatory head posture with good binocular function was discharged with binocular single vision and a normal head posture.

(*c*) *Palsy of Vertically-Acting Muscles with Secondary Convergence* (4 cases).—The following muscles were involved:

Superior rectus	3
Superior oblique	1

Two patients had symptoms, both with a compensatory head posture. One of these patients had good binocular function and the symptoms were relieved post-operatively with a return to a normal head posture. In the other case (a child aged 6 years) the deviation was constant pre-operatively

in spite of the presence of a compensatory head posture, but binocular single vision was restored after operation although the head posture remained abnormal.

The two patients who had no symptoms (aged 4 and 7 years respectively) both had a compensatory head posture and good binocular function pre-operatively. Full binocular single vision and a normal head posture were restored post-operatively.

SUMMARY AND CONCLUSION

It has been our aim in this article to stress the value of orthoptic treatment *in cases in which it is justified* and to emphasize that the time that is often spent in giving orthoptic treatment when it is not justified is time wasted. Great care should be taken in the selection of patients suitable for orthoptic treatment, particularly in the case of non-paralytic non-accommodative convergent strabismus.

The results in 213 cases of this type have been divided into three categories (Table III):

(1) Restoration of binocular single vision: 24 cases (11 per cent.)
(2) Subnormal binocular vision (equal or nearly equal visual acuity, normal retinal correspondence, doubtful fusion): 73 cases (39 per cent.)
(3) Defective binocular function (intractable amblyopia, lack of normal retinal correspondence, abnormal retinal correspondence, latent nystagmus): 116 cases (61 per cent.)

165 (77 per cent.) of these patients had orthoptic treatment (combined with operation in 138 cases) and the results correspond with those mentioned above:

(1) Restoration of binocular vision: 16 cases (10 per cent.)
(2) Subnormal binocular vision: 64 cases (39 per cent.)
(3) Defective binocular function: 85 cases (51 per cent.)

48 (23 per cent.) of these patients had no orthoptic treatment and in these results appear to be slightly better as far as Category 1 is concerned:

(1) Restoration of binocular single vision: 8 cases (17 per cent.)
(2) Subnormal binocular vision: 9 cases (19 per cent.)
(3) Defective binocular function: 31 cases (64 per cent.)

It is not suggested that a decision regarding the prognosis can always be made at the first visit, for in many instances, particularly in the case of young children, several diagnostic sessions may be necessary, but an early prognosis

concerning the anticipated standard the patient will reach in terms of binocular function should help to eliminate much unnecessary and unrewarding work and allow more time for concentrated treatment in deserving cases.

A study of the history in convergent strabismus shows that with few exceptions the results depend on the following factors:

(*a*) The age of the patient.
(*b*) The age at onset of strabismus.
(*c*) The mode of onset of the strabismus (*i.e.* constant from onset or intermittent).
(*d*) The duration of the strabismus before the visual axes are aligned by operation or other means.

Given the necessary data from the case history and from clinical investigation, and having decided upon the standard of binocular function which the patient is likely to regain, treatment can then be limited to achieving this end. Thus it should no longer be necessary to "try" orthoptic treatment. Orthoptic treatment is either indicated or it is not indicated. It is indicated only if there is a reasonable chance of thereby helping the patient to acquire some useful form of binocular vision. It is not indicated if there is clearly no chance of achieving any useful form of binocular vision.

The conclusions regarding prognosis and treatment that may be drawn from a study of the cases analysed in this article are summarized below.

PROGNOSIS AND TREATMENT INDICATED IN STRABISMUS
(With regard to binocular single vision)

(A) NON-PARALYTIC STRABISMUS

I. Convergent Strabismus

(1) Accommodative

(*a*) *Fully Accommodative.*—Prognosis, binocular single vision. Treatment, correction of refractive error, operation rarely needed.

(*b*) *Convergence Excess.*—Prognosis, binocular single vision. Treatment, correction of refractive error and in some cases operation.

(*c*) *Partially Accommodative.*—Prognosis, binocular single vision. Treatment of refractive error and operation.

Orthoptic treatment may be valuable in all types.

(2) Non-Accommodative

(*a*) *Of Early Onset*

(Birth to 18 months).—Rarely any chance of binocular single vision except in certain cases in which the visual axes have been made approximately parallel by operation within the first 2 or 2½ years of life.

(*b*) *Of Later Onset*

(2 to 3 years).—Binocular vision of a subnormal type may develop, provided the visual axes have been made approximately parallel, and orthoptic treatment may be of great value in obtaining it. Subnormal binocular vision may gradually be superseded by full binocular single vision.

(c) *Of Late Onset*

(3 to 5 years or later).—Normal binocular single vision should result following operation even if the latter is postponed until later in childhood or even early adult life provided adequate orthoptic supervision ensures equal visual acuity, absence of suppression etc.

Notes

(*i*) In non-accommodative convergent strabismus in which there is a vertical factor, the prognosis is no worse provided that the vertical deviation is adequately overcome by operation.

(*ii*) In any case of convergent strabismus in a child, provided there is good vision in each eye and provided that the angle of deviation is overcome by appropriate treatment, there is always a chance (even if a remote one) that some form of binocular vision may result.

(*iii*) If the strabismus is purely intermittent the prognosis is good.

II. Primary Divergent Strabismus

In general the prognosis is good even when the deviation is constant, (provided there is good visual acuity in each eye) for in many cases the strabismus has started in an intermittent fashion. Operation is needed in most cases but in some cases of intermittent divergent strabismus orthoptic treatment alone may suffice. The correction of the refractive error if myopic or astigmatic and the correction of anisometropia may be of great value.

(B) CONGENITAL PARALYTIC STRABISMUS

If binocular functions are good, even if binocular single vision is not demonstrable in any position of the gaze, the prognosis is good provided the surgical treatment is adequate. The existence of an abnormal head posture of the type that would be expected in the particular ocular palsy present indicates that binocular single vision has probably been present previously, and even if binocularity no longer exists the chance of its restoration by adequate surgical treatment (aided by orthoptic treatment) is good.

If binocular vision is absent the prognosis is usually poor unless surgical treatment is carried out at an early age.

In cases of strabismus where normal binocular function is thought to have existed previously (even if in rudimentary form) or is thought to exist potentially, the value of orthoptic treatment as a re-educating process is already known, but the application of our knowledge to the effects of disturbances of reflex development on the squinting child is still in the experimental stage. The restriction of orthoptic treatment to those cases in which there is a reasonable chance of improving binocular function would lessen much of the time-consuming work undertaken in an orthoptic clinic, and allow more time to be spent upon the detailed investigation and diagnosis of cases, especially those in which surgical treatment is required.

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KERATOCONJUNCTIVITIS SICCA DUE TO SARCOIDOSIS*

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SARCOIDOSIS is well recognized as a cause of swelling of the lacrimal glands and enters into the differential diagnosis of tumours of this region (Duke-Elder, 1952). Such swellings may be followed, in some cases, by a form of keratoconjunctivitis sicca (Ainslie and James, 1956; Gruber, 1956).

The association of systemic sarcoidosis with keratoconjunctivitis sicca in the absence of swelling of the lacrimal glands has also been described (Ainslie and James, 1956; James, 1956; Ramage and Kinnear, 1956), but in these cases the histology of the ocular lesions was not obtained; it was presumed, however, that they were of the same pathology.

In the case to be described, a patient, who had been treated in an ophthalmic clinic for 3 years for symptoms of conjunctival irritation, was found on mass radiography of the chest to have the typical radiological features of sarcoidosis. In view of this finding the patient was referred, after proof of systemic sarcoidosis had been obtained by liver biopsy, for investigation of her ocular condition. The patient's eyes lacked the generally accepted features of ocular sarcoidosis, namely swelling of the lacrimal glands, yellowish conjunctival follicles (Crick, Hoyle, and Mather, 1955; Crick, 1956), or chronic uveitis, but presented the clinical features of keratoconjunctivitis sicca. In addition she had complained of dryness of the nose and throat. Biopsy of lacrimal gland and conjunctiva, however, revealed that the ocular disorders were in fact due to sarcoidosis.

Case Report

A married woman aged 42, was admitted to The London Hospital for investigation in January, 1956. For 3 years she had complained of sticky threads forming in her eyes during the night. These were associated with a pricking sensation and discomfort which lessened during the day but worsened in the evening. For 3 years she had been treated for chronic conjunctivitis with a variety of drops, including penicillin and cortisone, without relief, and had been worsened by a course of silvering. She also noticed dryness of her nose and throat and would awaken with pharyngeal irritation and dryness, relieved by coughing up a tenacious plug of mucus.

For 2 years her chest had been under radiological observation since the discovery of hilar shadows and reticulation on mass radiography.

With the exception of intermittent pain in the right shoulder during the past 5 years,

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she had remained in good health until the onset of the ocular symptoms. Since then she had complained of lack of energy. She had one brother who died from tuberculosis.

Examination.—She was an athletic, well-nourished woman. Apart from the ocular features, the only abnormal finding was in the upper respiratory tract, where the nasal mucosa was relatively atrophic and rather dry. The mouth, larynx, and pharynx were normal. No abnormality of the skin, nails, salivary glands, lymphatic glands, or joints was discovered. The respiratory and cardio-vascular systems were clinically normal; blood pressure 130/90. The liver and spleen were not palpable and no abnormality was evident in the nervous system.

Laboratory Investigations

Chest x-Ray.—Diffuse infiltration in the lung fields especially in the upper lobes. Marked enlargement of the hilar lymph glands.

Sputum and Gastric Lavage.—No tubercle bacilli on smear or culture.

Mantoux Test.—Negative to 1 in 100 old tuberculin.

Erythrocyte Sedimentation Rate.—15 mm. in one hour.

Plasma Proteins.—Albumin/Globulin ratio 4·1/3·4.

Haemoglobin.—13·9 g. 100 ml.

Liver Biopsy (Fig. 1).—Needle biopsy showed non-caseous epithelioid cell tubercles with early peripheral fibrosis, morphologically compatible with sarcoidosis. No acid-fast bacilli.

FIG. 1.—Liver biopsy showing a non-caseating granuloma consisting of epithelioid cells surrounded by a narrow zone of lymphocytes. Haematoxylin and eosin × 150.

Ocular Examination.—The lids were normal but the palpebral conjunctiva showed a mild degree of chronic injection. There was a tenacious string of mucus in each lower fornix. A band of slight subepithelial infiltration stretched across each lower fornix (Fig. 2, opposite). This was thicker and yellowish in some areas and there were other slightly infiltrated areas, but the condition could hardly be regarded as follicular. The bulbar conjunctiva was normal in appearance apart from rather prominent yellowish pingueculae. Bengal rose stained the interpalpebral conjunctiva of each eye intensely

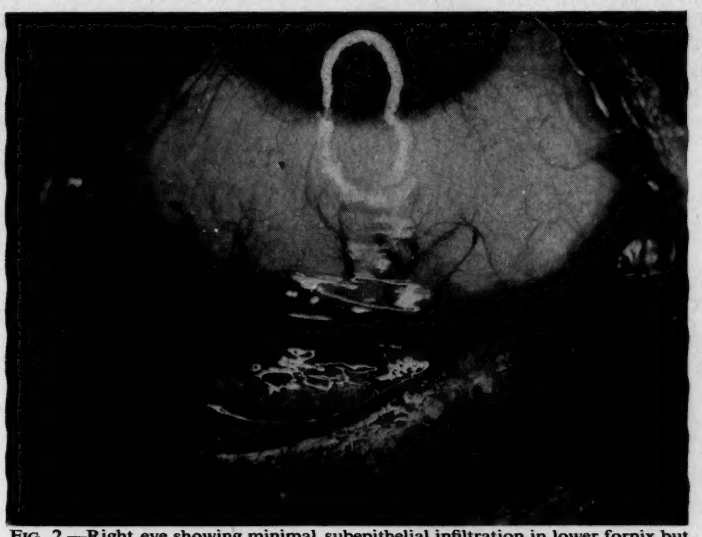

FIG. 2.—Right eye showing minimal subepithelial infiltration in lower fornix but none of the yellowish follicles usual in conjunctival sarcoidosis.

(Fig. 3), and with the Schirmer test only five mm. were wetted in 15 minutes in each eye. There was punctate staining of the lower third of each cornea with fluorescein or Bengal rose, but otherwise the globes were normal. Neither lacrimal gland could be palpated.

FIG. 3.—Left eye showing Bengal rose staining the interpalpebral conjunctiva.

Ocular Investigations.—No bacteria grew on conjunctival culture. Giemsa-stained conjunctival scrapings revealed no organisms and no virus inclusions in the epithelial cells. A scanty lymphocytic exudate was present and many of the epithelial cells showed the pink granulation of early keratinization.

Conjunctival biopsies from the infiltrated areas of each lower fornix and the left upper fornix were fixed in Zenker's fluid and stained with haemotoxylin and eosin. They showed an increased number of goblet cells and a slight subepithelial infiltration with lymphocytes and plasma cells (Figs 4 and 5).

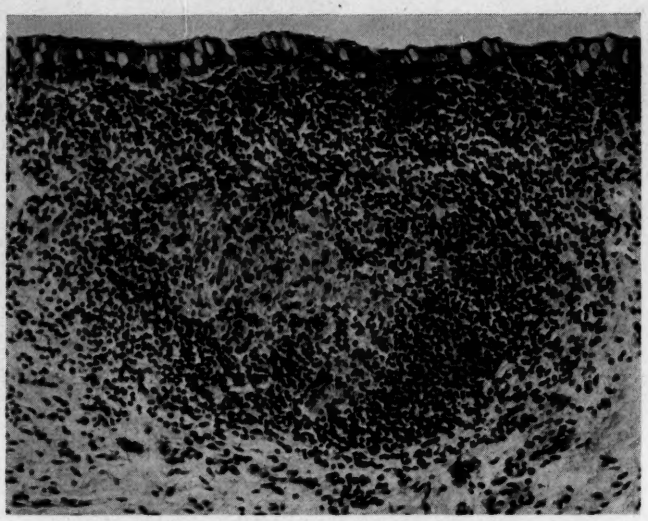

FIG. 4.—Subepithelial granuloma in conjunctiva from right lower fornix showing epithelioid cells without caseation surrounded by lymphocytes. Haematoxylin and eosin × 150.

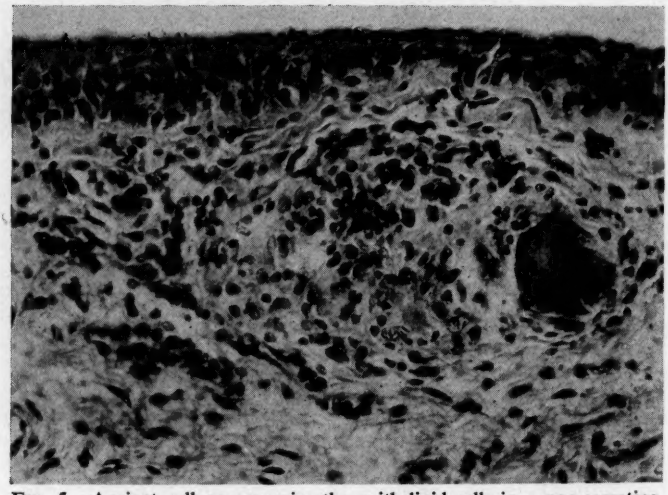

FIG. 5.—A giant cell accompanies the epithelioid cells in a non-caseating granuloma in conjunctiva of the left lower fornix. Haematoxylin and eosin × 330.

Small scattered subepithelial granulomata consisting of large eosinophilic epithelioid cells surrounded by small lymphocytes were seen, and giant cells of the Langhans type were present in several. No acid-fast bacteria could be demonstrated.

A biopsy of the lower pole of the left lacrimal gland (Fig. 6) showed a diffuse increase in the interstitial lymphoid tissue with large aggregations of epithelioid cells without caseation, in which no tubercle bacilli could be seen.

FIG. 6.—Lower pole of left lacrimal gland with overlying conjunctiva. Sheets of non-caseating epithelioid cells and a diffuse increase in the interstitial lymphoid tissue have largely disorganized this lobule. Haematoxylin and eosin × 53.

Treatment.—She was given cortisone 100 mg. daily orally, reduced after 3 months to 75 mg. daily, together with Izoniazid 300 mg. daily and para-aminosalicylic acid 3·5 mg. four times daily. She is continuing this treatment at the time of writing. There has been marked symptomatic improvement.

Discussion

Clinically this patient presented with the features of keratoconjunctivitis sicca and not those characteristic of ocular sarcoidosis. Moreover, the keratoconjunctivitis was associated with dryness of the nasopharynx, features more in keeping with Sjögren's syndrome. It was only when evidence of pulmonary sarcoidosis was brought to light that the possibility of a similar aetiology of the ocular condition was considered.

Sjögren's Syndrome.—Sjögren and many others (reviewed by Duke-Elder, 1952) have emphasized that keratoconjunctivitis most often arises without preceding ocular disease in middle-aged women and is often associated with polyarthritis, xerostomia, salivary gland enlargement, and rhinitis, pharyngitis and laryngitis sicca. Many other abnormalities have been recorded in patients with dry eyes (Duke-Elder, 1952) and added to the syndrome; in many cases the syndrome is incomplete. There is an unfortunate tendency for keratoconjunctivitis sicca and almost any associated feature to be diagnosed as "Sjögren's disease" without further systemic investigation or treatment.

Nature of Sjögren's Syndrome.—There are four main views of this syndrome. Firstly, it has been regarded as a disease in its own right which gives rise to some or many of the possible clinical features (Sjögren, 1933; Henderson, 1950; Coverdale, 1948, 1952; Cardell, and Gurling, 1954).

Secondly, the syndrome has been regarded as a manifestation or complication of rheumatoid arthritis. Thompson and Eadie (1956) recently supported this view by an extensive investigation of patients with rheumatoid arthritis, in which they established that keratoconjunctivitis sicca is the commonest ocular complication of rheumatoid arthritis. However, rheumatoid arthritis cannot be the aetiology of those fairly common cases of Sjögren's syndrome without arthritis unless they represent anarthritic rheumatoid disease (Bagratuni, 1956).

Thirdly, since keratoconjunctivitis sicca has been described in association with several collagen diseases other than rheumatoid arthritis, namely, rheumatic fever, polyarteritis nodosa, disseminated lupus erythematosis, and scleroderma, it has been suggested that Sjögren's syndrome is merely one variant of collagen disease (Ramage and Kinnear, 1956; *British Medical Journal* Editorial, 1956).

Fourthly, the syndrome may be regarded as a syndrome and no more, that is to say a symptom complex which may arise from a variety of pathological processes. Having recognized the syndrome in any particular case, we should attempt to make an aetiological diagnosis. Thorough systemic examination may establish the presence of one of the collagen diseases. It is at this point that sarcoidosis should also be considered, and the patient carefully investigated for systemic and ocular sarcoidosis. The present case clearly demonstrates the simplicity and value of conjunctival and lacrimal biopsy in making the diagnosis and in establishing that the systemic disease and the ocular condition are in fact due to the same cause.

From among patients with keratoconjunctivitis sicca some will also be recognized who are presenting the early stages of a disease resembling ocular pemphigus.

Features of Sjögren's Syndrome which can arise in Sarcoidosis.—The present case shows that keratoconjunctivitis sicca can result from lacrimal and conjunctival sarcoidosis even without swelling of the lacrimal gland, as described by Gruber (1956), or manifest conjunctival follicles as described by Crick and others (1955). Swelling of parotid and other salivary glands has been recorded in numerous cases of sarcoidosis (Longcope and Pierson, 1937; Pautrier, 1938; Scott, 1938; Schultz, 1945). Xerostomia would be expected in a patient whose salivary glands were infiltrated by sarcoid deposits or disorganized by subsequent fibrosis (James, 1956). The present patient also had symptoms of rhinitis and pharyngitis sicca and the nasal mucosa had a dry and somewhat atrophic appearance. Infiltration of the nasal, paranasal, pharyngeal, and laryngeal mucosa in sarcoid have been demonstrated (Poe, 1942; Bordley and Proctor, 1942; Wille, 1946). Enlarged superficial lymph nodes, spleen, and liver are all well-recognized occurrences in sarcoidosis. Like other possible features of Sjögren's syndrome, their significance varies with the presence or absence of arthritis. If a patient with keratoconjunctivitis sicca has rheumatoid arthritis and enlargement of lymph nodes and spleen, then she is probably presenting Felty's syndrome. If she has no arthritis then sarcoidosis is more likely. Mild pyrexia, raised erythrocyte sedimentation rate, and hyperglobulinaemia not uncommonly occur in sarcoidosis and are often found in cases of Sjögren's syndrome associated with rheumatoid arthritis.

Features of Sjögren's Syndrome not expected to arise from Sarcoidosis.—Polyarthritis of the rheumatoid type does not result from sarcoidosis, so that the presence of definite rheumatoid arthritis in any case makes sarcoidosis much less likely. James, Thomson, and Willcox (1956) have recently drawn attention to the occurrence of an acute polyarthritis in association with the erythema nodosum which may arise in sarcoidosis and this must not be confused with a true rheumatoid arthritis. The dry brownish-yellow longitudinally fissured nails described by Thompson and Eadie (1956) similarly appear to be a feature in those cases of Sjögren's syndrome associated with rheumatoid arthritis.

Summary

A case of keratoconjunctivitis sicca with symptoms of dryness at the back of the mouth was shown by biopsy to be due to lacrimal and conjunctival sarcoidosis. Systemic sarcoidosis was demonstrated by chest *x* ray and liver biopsy.

The relationship of Sjögren's syndrome to rheumatoid arthritis and to sarcoidosis is discussed, and it is suggested that general investigation, conjunctival biopsy and possibly lacrimal biopsy should be used to elucidate the aetiology in those patients who do not have a rheumatoid arthritis

accompanying their dry eyes. If the keratoconjunctivitis sicca is accompanied by yellowish conjunctival follicles, swelling of the lacrimal glands, or chronic uveitis, then sarcoidosis is more likely.

Although sarcoidosis may well account for only a small proportion of cases of keratoconjunctivitis sicca, these cases are worth recognizing since the disease is known to respond to cortisone and it is reasonable to suppose that if treatment can be maintained until it becomes inactive then permanent damage by fibrosis can be prevented.

We should like to express our indebtedness to Dr. Owen Clarke, Chest Physician of Canterbury, who suspected that both the eye and lung changes might be due to sarcoidosis and referred the patient to the London Hospital; also to Dr. D. Davies of Whitstable, the patient's General Practitioner, for his assistance in carrying out the treatment. We are indebted to Dr. N. Lloyd Rusby and Mr. J. E. M. Ayoub for their encouragement and permission to publish this case; to Dr. J. R. Pridie for the liver biopsy; to Dr. R. G. F. Parker and Dr. C. Wood for the histological reports; and to the photographic department of The London Hospital for the illustrations.

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Brit. J. Ophthal. (1957) **41**, 161.

MENINGIOMA INVADING THE OPTIC NERVE*

BY

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MENINGIOMATA of the optic nerve are rare tumours, although recently they appear to be somewhat less rare than is suggested in the earlier literature. Thus Mathewson (1930) could find only 33 reported cases, but Craig and Gogela (1949) were able to describe a single series of seventeen cases, nine arising within the orbit, three from the nerve within the optic foramen, and five apparently from other orbital structures. Reese (1951) found as many as 21 meningiomata (site not stated) in a series of 251 primary orbital tumours. Isolated cases are, therefore, hardly worthy of a separate report, but the one here described shows unusual and hitherto unrecorded features: the patient is the youngest yet reported, in only one other case was there a similar invasion of the optic nerve, and the number of ocular structures involved appears to be unique.

Case Report

A **boy aged 3** was brought to the Benn Eye Hospital on November 14, 1955, as his mother thought that his right eye had an inward squint, and that he could not see properly with it.

Examination.—It was found that he did not follow light with his right eye, and that there was a fibrous-looking mass behind the lens nasally.

Subsequent examination under general anaesthesia confirmed that there was a large white mass involving the nasal half of the fundus and obscuring the optic disc. The remainder of the retina was detached and a retinoblastoma was considered to be the most likely diagnosis. Enucleation was recommended but parental consent for operation was not at first forthcoming. Eventually enucleation was undertaken on January 4, 1956.

No evidence of extra-ocular tumour tissue was noted at the time of operation.

Further Investigations.—After the pathological report on the enucleated eyeball, full medical and neurological examinations were carried out, including *x* rays of skull, chest, and abdomen, but no abnormality was found.

Treatment.—Mr. A. R. Taylor, neurosurgeon, saw the patient and reported as follows:

"It is rather a difficult decision whether to subject the child to major neurosurgical procedures at the moment. Reading the pathological report carefully one cannot be absolutely certain that the whole tumour has not been removed. In any event we are losing nothing by waiting a little, because if the tumour recurs, it will not infiltrate into the cerebrum; it will form a mass which will remain removable. By keeping an eye on the orbit and *x*-raying the optic foramina at regular intervals, I think that we will have early notice of any recurrence."

No treatment other than enucleation of the right eyeball has been undertaken.

Present Condition.—He has remained well since and when seen in October, 1956, the right socket was healthy and no abnormality was noted in the media or fundus of the left eye.

X rays of the skull, including the optic foramina were normal.

* Received for publication December 3, 1956.

Observation.—It is proposed to repeat these examinations at regular intervals until we are reasonably certain that the tumour originated in the optic nerve and that it has been completely removed.

Pathological Findings.—The right eye was fixed in 10 per cent. formol saline.

Macroscopically the optic nerve measured 1·5 cm. from the globe to the cut end and showed no swelling. There was a funnel detachment of the retina with the inner layers in apposition, and on the naso-inferior side close to the disc the retina was swollen.

Microscopically there were two large masses of growth within the optic nerve close to the sclera immediately posterior to the lamina cribrosa. The larger mass had replaced the whole of the optic nerve at this point and had spread to the pia on one side, while the smaller mass surrounded the central retinal vessels and lay immediately posterior to the larger one (Figs 1 and 2).

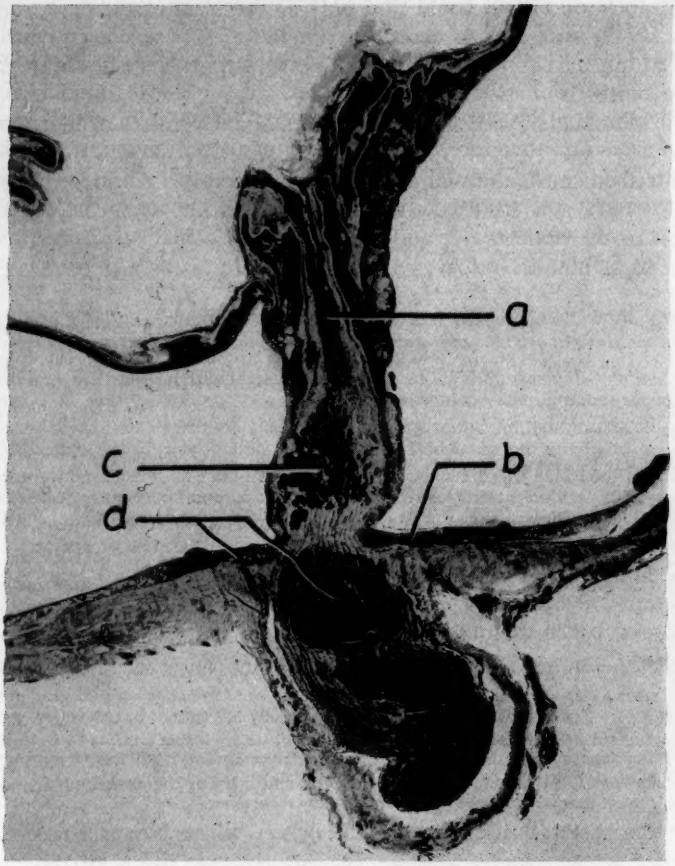

FIG. 1.—Posterior half of globe showing retinal detachment and meningiomatous invasion of:
(*a*) Inner layers of retina;
(*b*) Choroid on temporal side of disc;
(*c*) Optic nerve head;
(*d*) Choroid on nasal side of disc and optic nerve.
Haematoxylin and eosin × *10.*

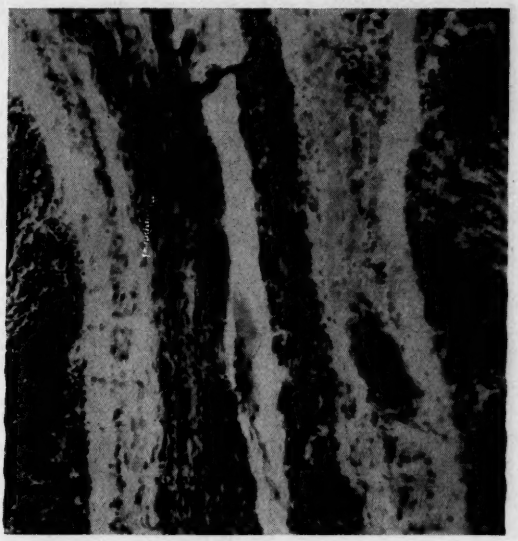

FIG. 2.—Opposing layers of detached retina invaded by meningiomatous growth. Haematoxylin and eosin ×90.

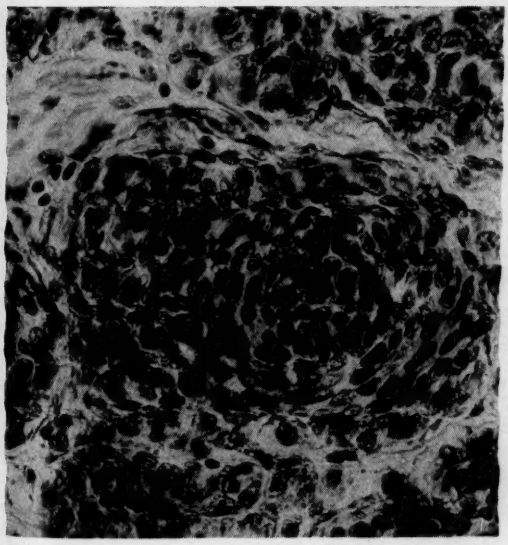

FIG. 3.—High-power view of optic nerve-head showing typical whorls of meningioma. Haematoxylin and eosin ×390.

Both were composed of densely-packed aggregations of large fusiform endothelial cells arranged in partial whorls segregated from each other by fine fibrous septa (Fig. 3). Tumour cells were present in the optic disc, in the adjacent choroid on both sides, and also in the stratum opticum of the detached retina. A transverse section of the cut end of the optic nerve showed no abnormality except for a collection in the extradural position of meningiomatous cells surrounding two psammoma bodies (Figs 4 and 5, overleaf).

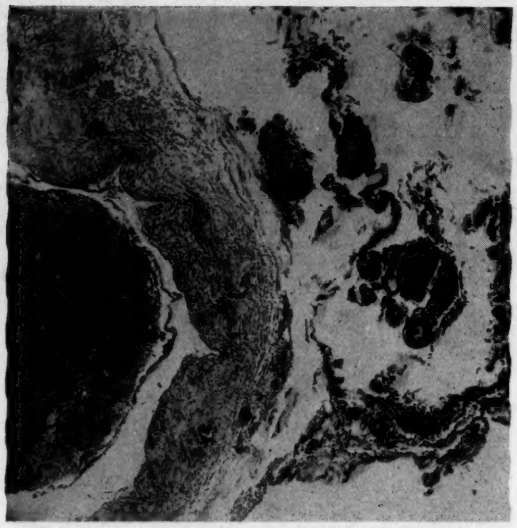

FIG. 4.—Cross-section of cut end of optic nerve showing normal nerve and sheaths with focus of meningiomatous cells and psammoma bodies extradurally. Haematoxylin and eosin × 36.

FIG. 5.—High-power view of extradural focus shown in Fig. 4. Note concentric laminated psammoma bodies. Haematoxylin and eosin × 195.

Discussion

Meningiomata of the optic nerve usually occur after the first decade and more than half of them after the age of 30 (Coston, 1936). In a collection of 29 cases, Hudson (1912) found that more than 50 per cent. were over 30 years of age, and in none of the remainder did the onset of symptoms occur before

the age of 6. Forrest (1947) gathered together a collection of seventeen cases in which the average age was 40.

The age of our patient is therefore of interest; it is the youngest yet recorded although the case reported by Ryan (1953) may have been even younger when the lesion first developed. Ryan's patient was a boy of 6 at the time of operation, but by this age the growth had advanced to such a degree that the eye was blind and fixed in the orbit, and the neoplasm could not be completely eradicated. Moreover, there was a history of squint since birth.

In view of the youth of our patient, it is remarkable that so many structures had been invaded, *i.e.*, the optic nerve, central retinal vessels, optic disc, choroid, and retina, and yet the growth within the sheath of the nerve and outside it was minimal. Involvement of so many sites has not been reported before, although there are reports of invasion of one or two of these structures. In the series reported by Hudson (1912), invasion had occurred in the choroid in four cases, in the choroid and sclera in three cases, and in the optic nerve in one case. Coston (1936) believed that optic nerve meningiomata show a pronounced tendency to invade the globe, and he described a case of invasion of the optic disc and retina with the nerve remaining free, although, as is usual, it was compressed into a thin cord. One of the seventeen cases of Forrest (1949) showed growth extending along the central retinal vessels of the optic nerve, and in another the retina was invaded at the disc.

Neame (1923) reported a case in which a meningioma was present in the subdural space and also within the atrophic retina near the disc margin, but made no mention of invasion of the optic nerve. A somewhat similar distribution occurred in the case of a boy of 14 described by Hope-Robertson (1949), in which the subdural space of the nerve and the posterior aspect of the eyeball were involved.

We have found only one reference to definite invasion of the optic nerve fibres by meningioma cells. This was reported by Brailey (1887), who described a sarcoma growing from the dural sheath of the optic nerve and occupying the central artery and the nerve itself. A beautiful etching of the tumour shows it to be a meningioma.

Casari (1947) classified meningioma of the optic nerve into three types:

(*i*) those growing from the sheath outwards,

(*ii*) those infiltrating the sheath,

(*iii*) those infiltrating the nerve.

However, he quoted no example of the third category, into which our case falls, and it is clear that, although invasion of the optic disc and retina may occur, extension into the optic nerve itself is extremely rare.

The problem of the exact site of origin of meningiomata has always excited controversy, and in the orbit, where a variety of differing structures

comes into close relationship, the question is particularly difficult. Indeed, it has been doubted whether meningiomata ever arise primarily within the orbit (Cushing and Eisenhart, 1938), and Verhoeff (1932), who on theoretical grounds accepted such an origin, had never seen a case. That they may do so, however, was well shown in a case of Friedenwald (1938), wherein a meningioma, together with the eye, was removed from the orbit; the proximal stump of the nerve adjacent to the foramen was normal, as also were *x* rays of this region, thus establishing the diagnosis of a primary intra-orbital meningioma.

The case here reported might be another such example, but the presence of tumour cells and psammoma bodies lying in the extradural region at the cut end of the optic nerve prevents a firm conclusion. It is possible that the tumour may have arisen intracranially or in the optic foramen, but there is no post-operative evidence of residual growth and the child is well.

We wish to acknowledge with gratitude the help and encouragement given to us by Dr. Norman Ashton during the preparation of this paper.

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ADDENDUM

While this paper was in the press, a publication by Shepard N. Dunn and Frank B. Walse appeared in the *A.M.A. Archives of Ophthalmology*, Vol. 54, No. 5, p. 702, in which a case of meningioma invading the optic nerve is described.

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INTRA-EPITHELIAL EPITHELIOMA*
BOWEN'S DISEASE

BY

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INTRA-EPITHELIAL EPITHELIOMATA may present to the ophthalmologist on the cornea, conjunctiva or eyelids, generally of elderly men. Here they form vascularized elevated plaques, that look rather gelatinous or granulomatous, and they most often arise in sites of chronic infection, as after burns or pannus. Years later they tend to break through the basement layer of the epithelium and finally yield distant metastases. To the ophthalmologist their interest and importance lies in the need for their early recognition, since in this precancerous stage they can so readily be excised with no ultimate danger to life or eye, and this deserves especial emphasis since they are generally flat and inconspicuous, and readily mimic a pterygium or localized lid infection.

In presenting here one further case, arising as a great rarity from the tarsal conjunctiva, it seemed a good opportunity to trace the subsequent history of those other seven cases that have so far reached the pathological department of the Institute of Ophthalmology, and so to assess the prognosis after the varied therapeutic approaches that have been used.

Case Reports

Case 1, a man aged 77, was referred to me by Dr. B. Bradley at the Central Middlesex Hospital for a rather irritable ulcer on the left lower tarsal conjunctiva; this was shallow and irregular, with a white heaped-up rim, about 10 mm. by 5 mm. in size, and spreading from the lid margin half way to the lower fornix (Fig. 1). A culture yielded only diphtheroids, and scraping showed no viruses or fungi. Treatment with various antibiotics produced little more than a variable lessening of the scant discharge and irritation.

FIG. 1. — Intra-epithelial epithelioma of lower tarsal conjunctiva (Case 1) before treatment.

* Received for publication December 12, 1956

On January 9, 1956, 6 months after the onset of symptoms, a biopsy was performed (Figs 2—6).

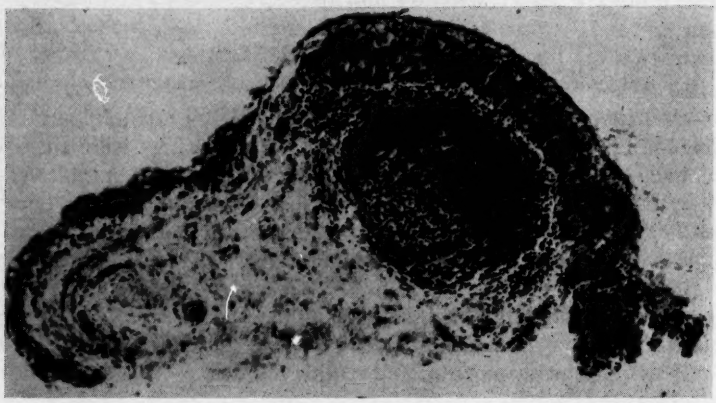


FIG. 2.—Case 1, general view of lesion, showing sharply demarcated area of epithelial hyperplasia with follicular inflammatory infiltration in subepithelial tissue. Haematoxylin and eosin × 67

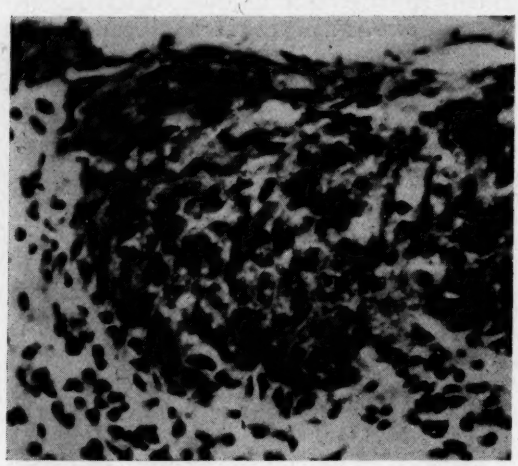


FIG. 3.—Case, 1, high-power view, showing loss of differentiation in epithelial layers. The cells are pleomorphic and show large hyperchromatic nuclei with numerous mitotic figures. Haematoxylin and eosin × 385

Dr. Norman Ashton reported as follows:

"Sections show a small piece of conjunctival tissue in which there is a sharply demarcated area of diffuse epithelial thickening involving two thirds of the epithelium. In consists of an undifferentiated mass of pleomorphic cells in active mitotic division and a few "monster cells" containing large hyperchromatic nuclei. There is no evidence of invasion of the subepithelial tissue which shows only diffuse and follicular infiltration with lymphocytes and plasma cells. The histological picture is typical of intra-epithelial epithelioma (Bowen's disease) of the conjunctiva."

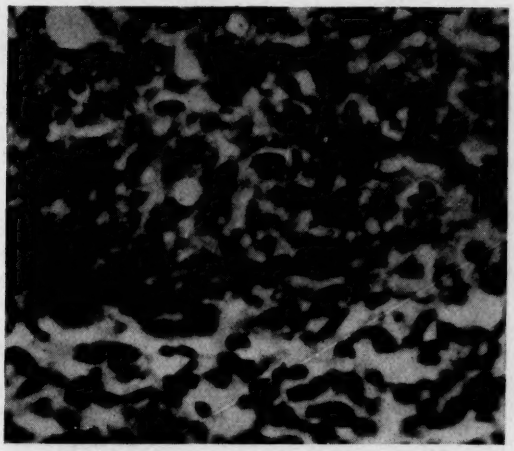
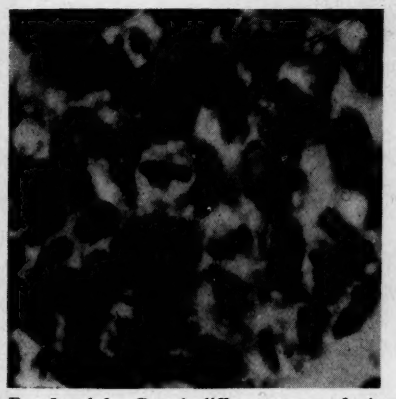
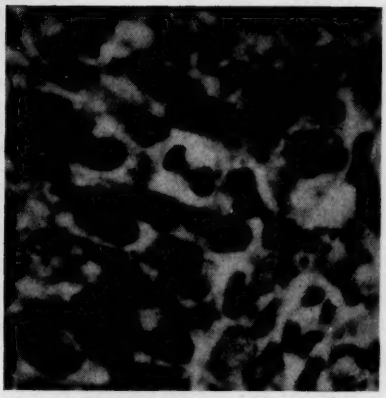


FIG. 4.—Case 1, high-power view of deeper epithelial layers showing haphazard arrangement of cells. Centrally a cell containing a single large nucleus ("monster cell") may be seen. Haematoxylin and eosin × 412





FIGS 5 and 6.—Case 1, different stages of mitosis in hyperplastic epithelium. Haematoxylin and eosin × 675

The affected area was then irradiated at Westminster Hospital, by closely applying to the everted lid a radio-active strontium shell (4,800 r, in six applications of 100 r/min. for 8 minutes at 2 to 3-day intervals). Within a fortnight the ulcer had completely dispersed (Fig. 7, overleaf) and has never recurred; the eye was already blind from cataract but the patient remains in other respects healthy.

Follow-up of Previous Cases from the Department of Pathology, at the Institute of Ophthalmology

Case 2, a man aged 46, had been attending the Cardiff Royal Infirmary under Mr. B. Gluck. For 8 years he had had a degenerative condition of the corneal epithelium in the form of raised white patches, some of which stained with fluorescein; and then 2 months previously had developed a greyish round patch astride the upper limbus 3 to 4 mm. in

FIG. 7.—Case 1 after treatment with β-radiation.

diameter, slightly raised, with many red spots which appeared to be blood vessels coming vertically to the surface (Fig. 8).

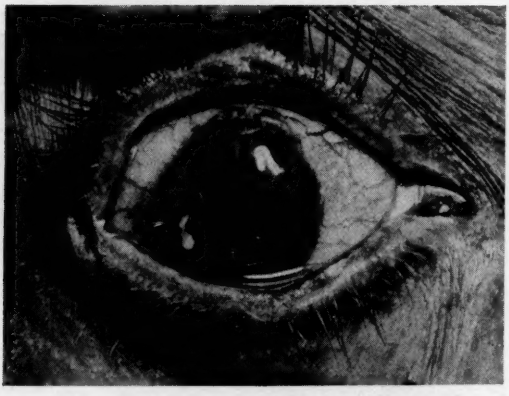


FIG. 8.—Typical intra-epithelial epithelioma of the limbus (Case 2).

A biopsy then showed the typical features of Bowen's disease, and the eye was enucleated on November 26, 1954; there has so far been no sign of any recurrence.

Case 3, a man aged 40, came from Johannesburg. His eye had been red for 6 months, with the development of a small semi-translucent lump (1 by 1·5 mm.) closely resembling a pterygium. This was excised, and, after reference through Sir Stewart Duke-Elder, was reported histologically as "Bowen's disease" (December 3, 1954). The eye has since then remained quiet, and there has been no sign of recurrence. (This was confirmed by patient and Dr. Frampton on November 15, 1956.)

Case 4, a woman aged 81, has already been reported by Greaves (1955). Her eye had been red and watery for 4 weeks, and exhibited a raised fleshy growth extending halfway across the cornea and well vascularized except over the part nearest to the corneal centre. A biopsy on December 15, 1954 showed the typical histology of "Bowen's disease". This was treated with superficial *x*-radiation 5000 r (400 r daily for 5 days a week). There had been no recurrence when the patient was last seen on January 1, 1956, and the vision had improved from 6/36 to 6/12, with no signs of an irradiation cataract.

Case 5, a man aged 54, was referred to Mr. C. Cook at Guy's Hospital with a localized gelatinous swelling at the limbus. This eye had been inflamed "off and on for years" and showed a "juicy swelling astride the limbus, with some corneal staining axially". After a few months' conservative trestment, a patch of xerosis developed at the limbus opposite the original growth, and then after a further few months, in which there was little change save an increasing corneal vascularization (with diminished sensation), the patient was admitted to hospital for biopsy on January 19, 1955. Both limbal swellings had the typical histology of Bowen's disease, whereupon both lumps were excised. A month later another tiny lump had arisen at the limbus midway between its two fore-runners, and this was also removed. The following month, persistence of the punctate epithelial erosions led to his reference to Dr. M. Lederman by whom the eye was irradiated (6240 r in four fortnightly applications of a strontium shell). Thereafter the cornea remained quiet but hazy and vascularized, and there had been no recurrence by January 1, 1956.

Case 6, a male Nigerian aged over 60, was seen by Dr. F. C. Rodger after 4 years of eye discomfort. A fleshy lump was found to cover the upper half of the cornea and adjacent conjunctiva, resembling a leprous sclero-keratitis; there was no associated trachoma. This was excised, and biopsy later confirmed the diagnosis of Bowen's disease. Unfortunately by that time the patient was frightened of any further measures, and had disappeared into the jungle without trace.

Case 7, a man aged 68, was under the care of Dr. R. D. Calcott of Nairobi. A limbal tumour had been removed on April 27, 1955, and the sections showed a squamous-cell carcinoma arising in a previously existing intra-epithelial epithelioma, with early invasion of the subepithelial tissues. The tumour was irradiated, and subsequently the eye remained quiet with no evidence of recurrence. However, by March 22, 1956, the dystrophic condition of the corneal epithelium had progressed with greater thickening and opacity of the cornea, although it was still clear enough at the axial area to allow 6/12 unaided vision. The patient then emigrated to Australia and has not yet been traced.

Case 8, a man aged 78, was referred to Mr. J. A. Magnus with a tumour at the limbus of the left eye that had been noted for the previous 14 weeks. This tumour was excised on December 7, 1955, apparently completely, and it was confirmed histologically to be an intra-epithelial epithelioma. No further treatment was needed and there has been no sign of recurrence.

Discussion

Bowen's disease is primarily one of a motley of precancerous conditions of the conjunctiva, well recognized since their initial description by Bowman (1849). Subsequent reports of such cases listed by Lugossy (1956) propose various names such as "tyloma", "hornification of the conjunctiva", "keratosis", "conjunctival callosity", and especially that of Lister and Hancock (1903), "epithelial plaque". These lesions have been classified clinically by Nicholls (1939) as:

(*a*) epithelial plaques with xerosis,
(*b*) congenital plaques,
(*c*) plaques secondary to chronic initiations (some of these last show a rapid hyperplasia similar to or identical with leukoplakia elsewhere.

They have been classified histologically by Janert (1956) as epithelial hyperplasia, without dyskeratosis, with dyskeratosis, and with hyperkeratosis (corresponding to the clinical pictures of erythroplasia, Bowen's disease, and leucoplakia). Although they may arise at any site on the cornea or conjunctiva, they usually appear at the limbus.

The clinical variant on the skin, described by Bowen (1912) and established as an important ophthalmological disease by McGavic (1952), has a very characteristic histological picture and is really a frank *carcinoma-in-situ* which has simply not yet spread from the epithelium of its origin. Thus it is proper to separate it from the various epithelial plaques of which the precancerous state can only be inferred by analogy, in many cases perhaps incorrectly. Microscopically the basal cells proliferate to form elongated pegs without actual invasion of Bowman's membrane; they show marked variation in size, shape, staining characteristics, and loss of polarity, some becoming "monster cells", containing either a single large nucleus (Fig. 4) or multiple small nuclei clumped together after amitosis. Some large cells contain a vacuole encircling the nucleus, others show premature keratosis, and mitotic figures are common (Figs 5 and 6). Beneath the epithelium there are signs of a mild chronic inflammatory reaction with lymphocytes, plasma cells, and histiocytes (Fig. 2).

The eight cases from the Department of Pathology at the Institute of Ophthalmology were all reported to show the characteristic histological features detailed in Case 1; with isolated exceptions they arose at the limbus, and occurred in men aged over 40. It is doubtless a curious coincidence that three of them should have come from different parts of Africa. In one case only had the cancerous change already begun. Multiple malignancy, noted in five out of sixteen cases reported by McGavic (1942) and in four out of eighteen reported by Locke (1956), was not recorded for any of these eight patients. It is interesting, however, that in Case 5 two foci arose at opposite sides of the limbus, and a third then followed after these had been excised. Pre-existing corneal or conjunctival disease (found in six of Locke's eighteen cases) was noted in none of the eight.

These eyes had all presented a red, irritable conjunctiva in which the diagnostic nodule (or ulcer, in the one eyelid to be affected) gradually appeared some months or even years later. They sometimes persisted as an indolent superficial keratitis with crops of punctate erosions, haze, and later vascularization, which gave little clue to the cancer that lay concealed.

Case 1 is of especial interest in that the lesion arose from the conjunctiva at the lid margin. All the other seven cases from the Institute of Ophthalmology and all of Locke's eighteen cases arose from the cornea or bulbar conjunctiva.

Treatment may be by excision of the tumour (Cases 3 and 8) and this may even entail removal of the whole eye (Case 2) by irradiation (Cases 1 and 4), or by local excision followed by eradication (Cases 5 and 7). Small tumours

could doubtless be expunged with equal certainty, especially from the lid, by cauterizing chemicals or diathermy. In only one of our eight cases (Case 5) has the lesion definitely recurred although at a different section of the limbus. The corneal opacity has progressed only in Case 7. No further records were available from Case 6.

In his follow-up analysis of eighteen cases, Locke noted recurrence in three out of ten excised locally (re-operation being successful in all three) and in one of five more severe cases that had required enucleation (in this one the recurrence was by direct extension to the lacrimal sac). Three of Locke's cases had been treated by *x*-radiation, one was unsuccessful (dosage not known), and in two advanced cases where the growth was destroyed, one had a parotid metastasis, and the other lost the eye from iritis and secondary glaucoma.

The length of the follow-up period in the foregoing eight cases ranges from 1 to 2 years. This is too short for any mature conclusions to be drawn but, since the site of the tumours lends itself to such minute examination, it would be reasonable to expect by now some recrudescence to be manifest in any tumour that had been inadequately despatched.

For epibulbar tumours the choice between excision and irradiation may well be dictated by the size of the growth, the extent of the damage to sight, and the availability of *x*-radiation, but local excision is probably the simplest and surest method. The dose of *x*-radiation recommended in each case was fairly heavy, but such a dose can apparently be tolerated well by the eye.

For the rarer tumour on the palpebral conjunctiva, where distance from the eyeball renders any procedure relatively safe, daily contact-applications of the strontium shell in the out-patients' department may well be the simplest solution.

My acknowledgements are due to Dr. Norman Ashton, Director of the Department of Pathology at the Institute of Ophthalmology, for his considerable assistance in the pathological assessment and in preparing the photomicrographs, to Dr. Peter Hansell of the Department of Medical Illustration, Westminster Hospital, for the illustrations of Case 1, and to Mr. B. Gluck for permitting me to use his photograph of Case 2 from the Medical Illustration Department, Cardiff, Royal Infirmary. I am also indebted to the several surgeons in charge of the other six cases for their helpful information: Sir Stewart Duke-Elder, Dr. G. Frampton, Mr. D. P. Greaves, Mr. C. Cook, Dr. F. C. Rodger, Dr. R. D. Calcott, and Mr. J. A. Magnus.

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A METHOD FOR THE DIRECT OBSERVATION OF RETINAL VESSELS IN THE EXPERIMENTAL ANIMAL*

BY

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IN the original studies of Ashton and his co-workers on the effect of high ambient oxygen concentrations on the blood vessels of the immature retina (Ashton, Ward, and Serpell, 1953, 1954), a method was devised whereby it was possible to observe the retinal vessels of an anaesthetized animal through a stereoscopic microscope (Ashton and Cook, 1954). This technique, primarily designed for work in connexion with the aetiology of retrolental fibroplasia, clearly has many other applications both within and beyond the field of ophthalmological research, since it provides a fairly straightforward means of directly observing and correlating retinal vascular changes with other physiological data.

The apparatus used for this work has now been modified and developed; many of the earlier technical difficulties have been overcome and the new methods have proved entirely satisfactory in numerous experiments carried out during the past year and in the preparation of a film. We have, therefore, decided to publish a detailed account of the apparatus and technique at present in use in this laboratory.

Apparatus

Fig. 1 (opposite) shows that the main frame is constructed from steel scaffold tubing, which properly stressed provides an immensely rigid framework and easily supports all the main units. These comprise the lighting system, animal chamber, microscope, ciné camera, and control gear.

Lighting System.—The source used (B, Fig. 2, overleaf) is a 4-mm. Tungsten arc of very high intensity surrounded by a steel box, which, although primarily acting as a screen for stray ultra-violet irradiation, also affords some protection in case of a tube explosion, for, although we have never experienced this, it is a distinct possibility, and the arc should never be mounted in an unshielded position. The arc housing is fixed coaxially to a telescope tube, which contains a heat-absorbing glass, paired condensers, and a final biconvex lens. The convergent beam of light which emerges from this is brought to a point of focus approximately 1 cm. behind the reflecting surface of the plane mirror (B, Fig. 3, overleaf). It is then reflected downwards through a right angle past the glass of the eye chamber (C, Fig. 3) and onto the retina (D, Fig. 3). The diameter of the spot of light thus produced in the eye can be varied by the iris diaphragm built into the lens tube.

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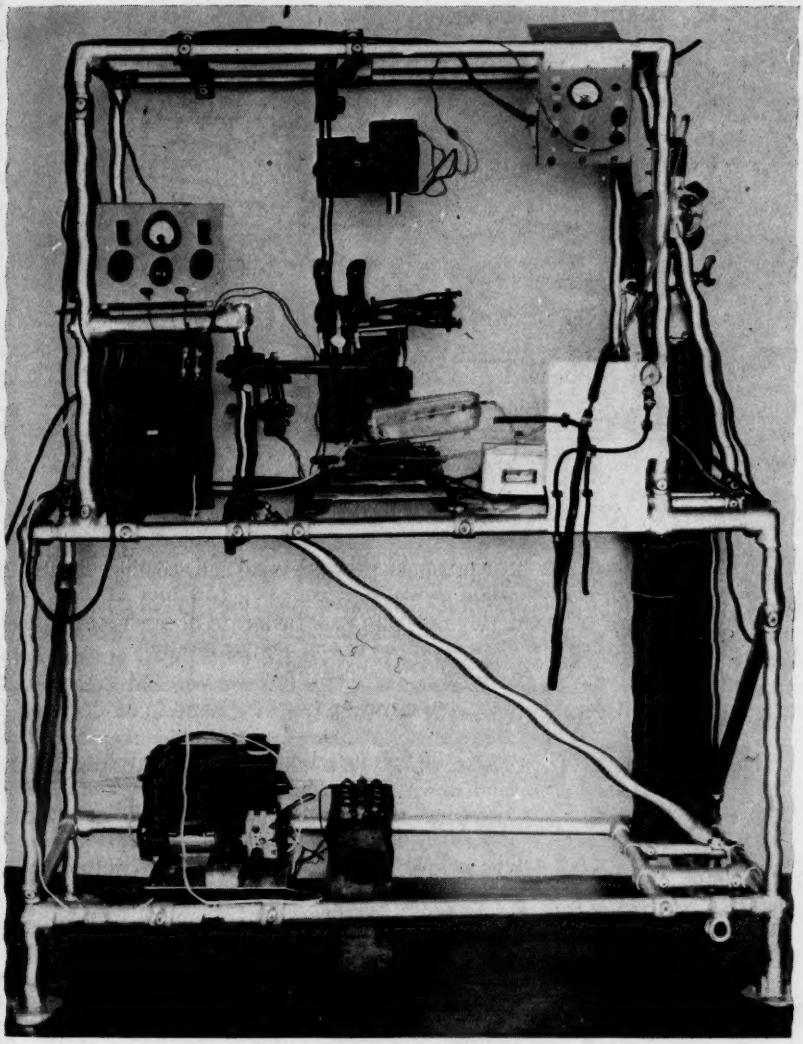

FIG. 1.—General view of apparatus.

Animal Chamber (Fig. 4, overleaf).—This is made from a commercially obtainable plastic food container, and is essentially a rectangular box with an airtight lid divided transversely, so that the section over the animal's head can be removed separately. This part also has a small hole cut in its centre to coincide with the position of the rim of the eye chamber (*vide infra*). On opposite sides of the box there are two tube connexions, one connected to the oxygen supply via a flowmeter and reduction valve, and the other leading to a B.O.C. oxygen analyser. This device

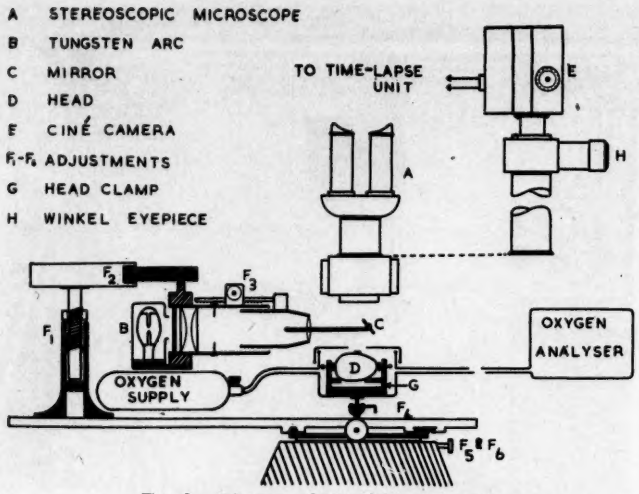

FIG. 2.—Diagram of essential components.

which operates on a diamagnetic principle, provides rapid and accurate readings of the ambient oxygen concentration inside the chamber. The floor of the box is attached to the base plate carrying the lamp unit, by means of a self-locking universal joint so that it can be tilted and fixed in any oblique position.

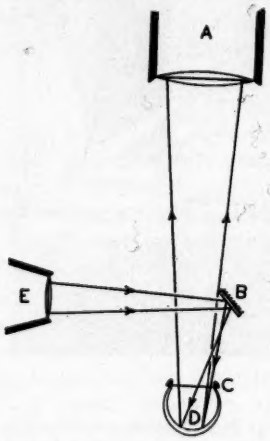

FIG. 3.—To show method of illumination.

Microscope.—This is a conventional Zeiss stereoscopic microscope (A, Fig. 2 and 3) mounted on a horizontally-swinging arm suspended from the main frame, so that its action together with the normal up and down movement of the focusing adjustment provides the necessary universal movement. Normally, a binocular head is used, but this is easily detachable, and can be quickly replaced by the Winkel eyepiece and ciné camera (E and H, Fig. 2).

Ciné Camera.—The first camera to be used was a Ciné Kodak special, fixed to the main frame so that it operated over one eyepiece of the binocular head. This was fitted with an electrically-operated shutter release mechanism, which in turn was connected to a simple electromechanical time lapse unit. Using this linkage, a shutter speed of about one frame every 2–6 sec. was used over periods of up to half an hour.

A second camera unit has now been built, consisting of a modified G.S.A.W. Gun Camera fixed to a Winkel eyepiece which fits into the base of the stereoscopic microscope.

Controls.—The adjustments necessary for the correct positioning of the components have already been mentioned, and are in fact largely self-explanatory on

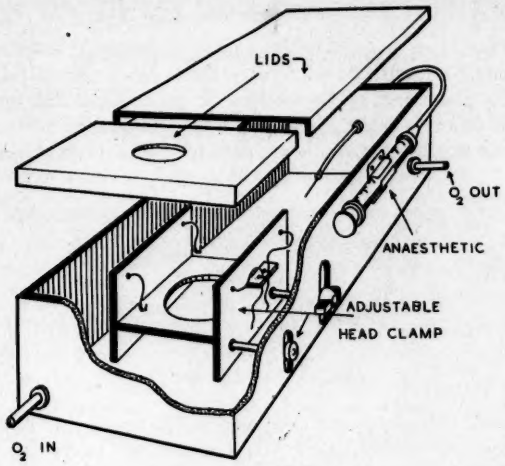

FIG. 4.—Animal chamber.

reference to Fig. 2. The only part requiring a separate note is the time lapse unit for the camera. Essentially, this consists of a motor-driven gearbox (F, Fig. 5) with an intermittent final drive rotating the disc (G, Fig. 5) through one quarter of a revolution per second. On the disc are mounted four projecting rods, three of which are of different length, so that when the disc rotates, the contacts J, which lead to the camera solenoid, are alternately made and broken. Thus if a different camera speed is required, the screws (L) are loosened and the contacts moved away from, or nearer to the disc, causing a different number of projecting pegs to strike the shutter contacts as the disc rotates.

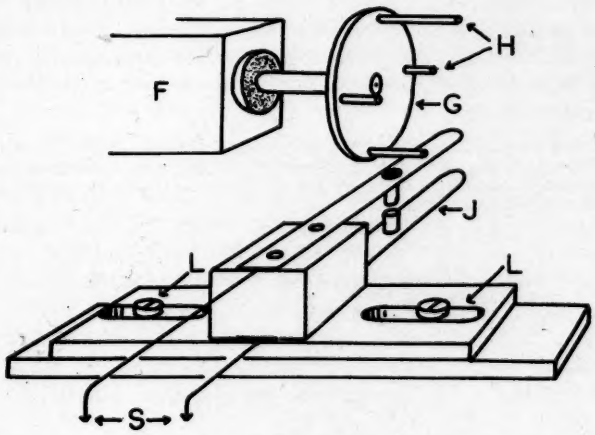

FIG. 5.—Time lapse unit.

Technique

Before the animal can be fixed in the oxygen chamber, it is necessary to remove the anterior ocular structures and replace them by a cylindrical metal chamber containing a flat glass disc. The method of performing this operation has not changed since it was first devised and the only modification which has been made is in the design of the "limbal window", details of which can be seen in Fig. 6.

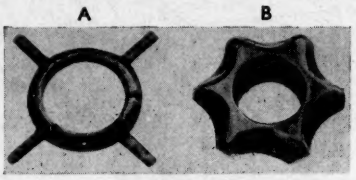

FIG. 6.—Limbal window.

After removal of the eyelids and conjunctiva, the brass ring (A, Fig. 6) is secured to the limbus by four sutures inserted through holes in its rim. The cornea is then removed with a cutting diathermy knife and the lens extracted, leaving behind the posterior part of the capsule as a temporary means of retaining the vitreous. Finally the inner chamber (B, Fig. 6) is screwed into the outer ring and the glass on its under surface allowed to impinge on the anterior vitreous face. In this way, the pressure exerted on the remaining intra-ocular contents may be varied within fairly wide limits, for if intra-ocular pressure is not maintained, then the retina will detach completely within a short time.

Once this "limbal window" operation is completed, the animal is transferred to the plastic chamber, and its head fixed rigidly by the clamps (G, Fig. 2). A needle is then inserted intraperitoneally and connected *via* a short length of polythene tube to a Mantoux syringe clipped to the outside of the box. This provides a convenient way of administering further doses of anaesthetic without disturbing the animal or the apparatus. Finally the airtight lid already referred to is applied and relative positions of the mirror, box, and microscope altered until optimum illumination of the retina is obtained. In this way the retinal vessels can be studied for as long as 10 hrs continuously at magnifications varying between 26 and 66 times.

I am most grateful to Dr. Norman Ashton for his help and encouragement during the preparation of this paper, and also to Dr. Peter Hansell for his help with the photographic apparatus described. I should also like to thank Miss E. FitzGerald for clerical assistance and Mr. G. Knight for his valuable technical help.

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STUDIES ON DEVELOPING RETINAL VESSELS*

IV. EFFECT OF IONIZING RADIATION

BY

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THE EFFECT of hyperoxia on the vasculature of the immature retina is now well established, and it has been repeatedly demonstrated that when a newly-born kitten is subjected to an ambient oxygen concentration exceeding approximately 35 per cent. a total and permanent vaso-obliteration will develop within a period of 8 to 36 hours (Ashton, Ward, and Serpell, 1954). This observation has provided the basis for much of the significant work carried out on the aetiology of retrolental fibroplasia, and may in fact provide useful information about other diseases of the retina. In view of this, it is clearly essential to assemble more information about the mechanism by which oxygen is able to produce vaso-obliteration in the immature retina.

In recent years a considerable amount of work has been published suggesting that the toxic effects of both oxygen and ionizing irradiation may be brought about by the inhibition or destruction of common enzymatic systems. For example, reduction of metabolism in the experimental animal by cooling, provides some protection again oxygen poisoning, whereas raising the body temperature enhances oxygen susceptibility (Grossman and Penrod, 1949; Stadie, Riggs, and Havgaard, 1944). Similarly it appears that a lowered metabolism decreases the amount of damage incurred by a given dose of radiation (Ord and Stocken, 1953). Furthermore, both agents affect some of the enzymes of intermediary metabolism in a comparable fashion; for instance, the sulphydryl enzymes, pyruvic oxidase and succinate dehydrogenase, are both inactivated by high oxygen concentration (Dickens, 1946), and the activity of diphosphoglyceric-aldehyde dehydrogenase is almost completely destroyed *in vitro* by a radiation exposure of 500 r. (Barron, Dickman, Muntz, and Singer, 1949). These reactions are very probably produced by the interaction of various oxidizing free-radicals transiently elaborated by ionising radiation from the breakdown of water, for there is considerable evidence to show that irradiation acts indirectly by forming toxic peroxide and hydroperoxyl ions (Rajewsky, 1952; Dainton, 1951).

In fact the yield of these substances is very considerably enhanced by increasing the quantities of molecular oxygen present; indeed, the incidence of chromosomal aberrations in inflorescences of algae can be made to rise as the oxygen concentration increases (Giles and Riley, 1950), and certain animal tumours can be made as much as three times more radiosensitive if they are irradiated in atmospheres of high oxygen concentration (Gray,

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Conger, Ebert, Hornsey, and Scott, 1953). Conversely, it has been shown that an anoxia produced by breathing 5 per cent. oxygen for 8 minutes protected rats from otherwise lethal doses of whole body radiation (Dowdy, Bennet, and Chastain, 1950). These facts have been applied to the treatment of human malignant disease with some success (Churchill-Davidson, Sanger, and Thomlinson, 1955; Hultborn and Forssberg, 1954).

Thus ionizing radiation and oxygen are apparently synergistic in their toxicity and both agents almost certainly act through similar enzyme systems. These arguments form the basis of the following experiments which were carried out to determine whether irradiation of the immature retina might parallel the vaso-obliterative effects of oxygen.

Technique

The irradiation source in these experiments was a cylinder of Cobalt 60, 4 mm. in diameter and 4 mm. in length. Its gamma equivalent was 1·4 milli-electron volts and its half life 5·3 years. The cylinder which was screened to emit gamma rays only, was enclosed in a lead bomb in such a way as to produce a surface contact dose rate of 240r/hr and a 5-mm. rate of 100r/hr.

In the first group (Table, opposite), each of the five animals was exposed to a different dose of irradiation, killed at once, and the retinal blood vessels injected with Indian ink. In the second group, four animals were exposed to the same dose of irradiation but kept for much longer periods before being killed and injected. In the third group, an attempt was made to watch the vessels whilst irradiation was in progress. For this it was necessary to modify the apparatus we had constructed for the direct observation of retinal vessels. The cobalt container was mounted on a swinging arm above a perspex box containing the kitten, so that it could be moved away and lowered into a protective lead surround in a very few seconds. The Zeiss stereoscopic microscope used for these observations was also mounted on a rotating arm, so that, when the cobalt was swung away into its safe position, the microscope could be moved very rapidly into place over the animal to observe the retina. In this way it was possible to see the retinal vessels at regular intervals throughout the irradiation. Before the commencement of the experiment, the kitten was anaesthetized with Nembutal and a limbal window inserted into the front of one eye using the same technique as that described by Ashton and Cook (1954). The head of the animal was then fixed in a rigid clamp, so that the radiation beam passed through one eye only, the other eye acting as a control and receiving approximately 10 per cent. of the total dose administered.

Results

In Group I, only one animal (k1) showed any abnormal vessel pattern after injection. In this case there was no injection of the vessels on the temporal half of the retina; however, it was impossible to decide whether this disappearance was due to the irradiation or to a thrombus in one of the main arteries.

In Group II, the long-term survival experiments, the vessels of both control and irradiated eyes appeared very similar in all animals.

In Group III, the size and general appearance of the vessels were recorded during the actual irradiation. In one animal (k10) the retinal arteries and veins were seen to become grossly dilated, but this was thought to be a result of its terminal condition. In the other two animals, no change at all was observed in the retinal vasculature during the course of the irradiation.

TABLE

Animal No.	Age (days)	Dosage Rate (r/hr)	Total Dose (r)	Estimated Control Eye Dose (r)	Survival Time (hrs)	Group	Experiment
k1	6	100	50	5	½	I	*Short Survival*
k2	8	100	100	10	1		Killed and injected with Indian ink
k3	8	240	480	48	24		
k4	8	240	960	96	8½		
k5	9	240	960	96	4½		
k6	13	240	960	96	7 days	II	*Long Survival*
k7	12	240	960	96	10 days		Killed and injected with Indian ink
k8	7	240	960	96	21 days		
k9	10	240	180	18	6½	III	*Direct Observation*
k10	13	100	500	50	7½		
k11	13	240	1200	120	2½		

The results of these experiments were completely negative. That is to say, we found no consistent change in the appearance or calibre of the vessels as a result of the irradiation doses employed. Any change that was observed was fairly easily explicable on the grounds of a variation in one or more of the experimental conditions.

Thus, these experiments provide no evidence to suggest that the biochemical systems known to be affected by both oxygen and irradiation are essentially involved in the vaso-obliterative effects of oxygen.

I should like to express my gratitude to Dr. Norman Ashton for his invaluable help and advice; also to Dr. Wilson, of the Physics Department, Westminster Hospital, for supplying the radioactive cobalt.

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TWENTY-SIX CASES OF EXTRA-CONJUNCTIVAL IMPLANTS*

BY

M. L. NAIRAC

Sunderland

TWO YEARS AGO I described a new implant so designed as to have the minimum of foreign material buried under the conjunctiva, and made the claim that because of this characteristic it could not be extruded (Nairac, 1954). During the period April, 1953, to July, 1956, 26 of these implants have been fitted, and not one has been extruded. The majority of cases have good movement and clean sockets. The Table shows an analysis of the 26 cases.

TABLE

RESULTS IN 26 PATIENTS

Case No.	Age	Date of Operation	Complications	Results
1	69	20.4.53	—	Good Clean socket Some limitation of movement of inferior rectus
2	15	24.6.53	Superior rectus cut out soon after	Repaired successfully Good result when last seen 12 months ago
3	57	13.8.53	Superior rectus cut out	Patient refused further operation Implant still in, other muscles holding well Limitation of elevation
4	30	27.8.53	—	Movement not full, but adequate
5	2½	1.10.53	Infection after measles *Inferior rectus cut out* soon after	Granulation tissue gave trouble for 3 months, *when clear, inferior rectus resutured* Still some discharge Fair movement
6	29	12.11.53	Inferior rectus cut out on 9.1.54	Inferior rectus resutured 22.11.54 Now good movement, clean socket Muscles holding
7	10	13.1.54	*Granulations over inferior rectus*	*Cleared in 3 months* Still a little discharge Fair movement
8	26	20.1.54	Inferior rectus soon cut out	Inferior rectus resutured 10.2.54 Good result, full movement, clean socket

(*contin.*)

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TABLE (*contin.*)

Case No.	Age	Date of Operation	Complications	Results
9	16	8.3.54	Further trauma March, 1955	After several attempts at repair, implant removed in February, 1956, because of persistent infection
10	10	17.5.54	Inferior rectus cut out 6.6.54	Inferior rectus resutured 26.7.54 Good result and movement
11	22	30.8.54	—	Some limitation of movement Clean socket
12	18	21.9.54	—	Good movement Clean socket
13	4	13.10.54	Granulation	Still some discharge, but no granulation Good movement
14	20	8.11.54	—	Good
15	52	14.12.54	—	Fair Some limitation of movement
16	46	27.1.55	—	Good
17	10	9.4.55	—	Good movement Clean socket
18	28	27.4.55	—	Good movement Slight discharge
19	11	6.6.55	—	Good
20	21	10.6.55	Granulation	Slight discharge until granulation excised 2.10.56 Socket now clean Good movement
21	41	27.6.55	—	Good
22	2½	17.11.55	—	Slight discharge, which will clear Fair movement
32	8	28.11.55	—	Good
24	34	16.1.56	—	? (moved out of district)
25	33	22.3.56	Threatened cellulitis 9.6.56	Cleared quickly with penicillin injections Socket now clean Fair movement
26	40	7.6.56	—	Some discharge, which will clear Good movement

In one case the implant has had to be *removed* because of intractable granulation tissue formation and threatened contraction, after a further injury 12 months later, when the internal rectus was torn from its bracket.

Attempted repair proved unsuccessful and infection prevailed. I stress that *this implant was removed, not extruded.*

An occasional muscle has cut out soon after operation, but has been successfully re-attached. This happens usually when the muscle has been stitched round the bracket either too tightly or inadequately.

To obviate this occasional muscle cut-out a new operative technique has been evolved which gives very satisfactory results.

Instead of attaching each muscle to its corresponding bracket as was done previously (Fig. 1), opposite muscles after passing through their individual brackets are attached to each other behind the implant and under the conjunctiva (Fig. 2). After enucleation of the globe, the threading of the four recti through their respective conjunctival slits, and the suturing of the central conjunctival hole (Fig. 3), the superior rectus is passed through its corresponding bracket of the implant from outside in, threaded back through its own conjunctival slit, and passed behind the conjunctiva and out through the conjunctival slit of the inferior rectus. It is then threaded through the bracket corresponding to the inferior rectus from inside out, and stitched firmly with catgut to the inferior rectus muscle. Fig. 4 shows the positioning and suturing of the muscles. The internal and external recti are then dealt with in a similar manner. The portion of the bracket covered by

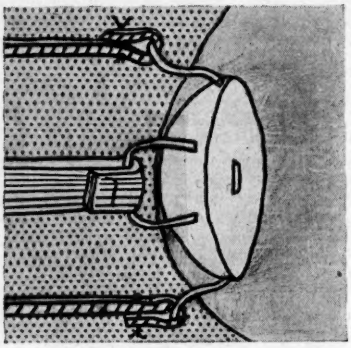

FIG. 1.—Old technique (Nairac, 1954; Fig. 4).

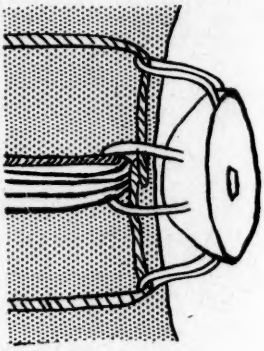

FIG. 2.—New technique.

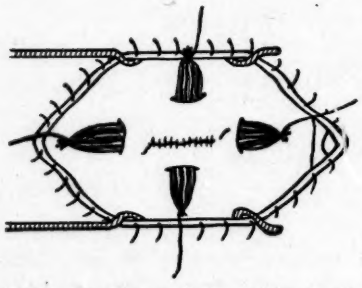

FIG. 3.—The four recti drawn through slits in the conjunctiva (Nairac, 1954; Fig. 3).

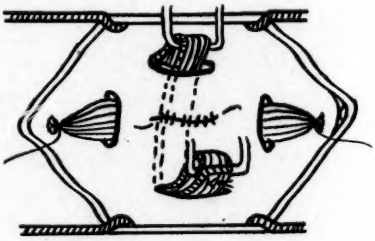

FIG. 4.—Method of suturing opposite muscles.

muscle is next allowed to slip back through the slits, and the four slits are carefully sutured over the muscles.

The claims that I make for this type of implant are that it is simple, affords better mobility than any type of totally buried implant, lies in a usually clean socket when simple hygiene is applied, can be used on patients of any age, and will remain *in situ* even when one muscle cuts out (Case 3). There is no evidence after 3½ years of any thinning of the tissues over the embedded bracket, and this series more than disposes of the criticisms that extrusion would occur within "1 to 2 years at the most" (Choyce, 1954), ulceration would occur at the point of entry of the brackets (Arruga, 1955).

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Brit. J. Ophthal. (1957) **41**, 186.

HEREDITY OF GREEN-BLINDNESS*

BY

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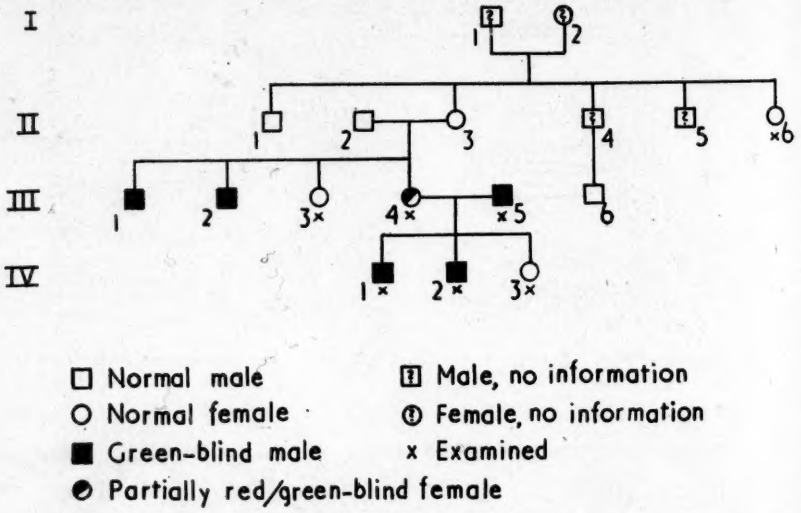
MARRIAGE between members of two families, in each of which red/green-blindness occurs, is rare. The publication of such pedigrees seems justified if only to confirm suppositions and hypotheses which have already been put forward.

Certain members of a family whose pedigree is shown in the Figure have been examined with Ishihara's Tables (1951). The persons tested are designated in the Figure as II, 6; III, 3; III, 4; III, 5; IV, 1; IV, 2; IV, 3.



Information was also collected about other members of the family. II, 1 and II, 2 were naval officers, and III, 6 was also in the navy, but died before achieving commissioned rank. III, 1 and III, 2, who are medically qualified, state that they suffer from the same anomaly as their nephews, IV, 1 and IV, 2.

Of the persons examined, III, 5, IV, 1, and IV, 2 are undoubtedly green-

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blind. The female patient III, 4, who read Ishihara's Tables as follows, gave the impression of a partial red/green anomaly:

Normal, eighteen
As with a red/green anomaly, fifteen
Doubtful, four

It could not be decided from these tests whether she showed a protanomaly or a deuteranomaly, since she read both figures in the critical Tables (Ishihara 24 and 25) correctly. However, her brothers and her sons are green-blind, and a green anomaly is therefore likely.

III, 4 and her daughter, IV, 3 are particularly interesting. The mother must be heterozygotic, otherwise the entirely normal colour perception of the daughter (IV, 3) cannot be accounted for.

The pedigree shows that heterozygotic carriers may present with pronounced abnormalities of colour perception, or may be phenotypically normal, both types of carrier occurring in the same family.

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Brit. J. Ophthal. (1957) **41**, 188.

THE ZAGORA ROD TEST*

BY

EDWARD ZAGORA

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CERTAIN mechanical operations in industry are possible only to persons with visual perception of the highest order. These high visual standards are of paramount importance to maximum production and to the prevention of occupational injuries. The practically simultaneous perception of depth and height is of special interest, for although such perceptions have a physiological basis, they are not innate, and some people with high visual acuity in each eye and normal fusion, have poor depth and height perception and cannot readily estimate distances.

Apparatus and Procedure

This test belongs to the rod-tests since the objects compared are three vertical rods each 10 mm. in diameter, two being stationary and the third movable. The apparatus stands on a table at eye-level (Figure). The first fixed rod is set at a distance of 6 m. from the subject and is placed 3 cm. to the right of the mid-line. The second is placed 3 cm. to the left of the mid-line and 6 cm. further away from the subject. Above the two stationary rods is suspended a movable one. All three rods are dull black and must have the same reflectivity. They are observed through an aperture 20 cm. wide and 12 cm. high in a screen placed 30 cm. in front of the first fixed rod.

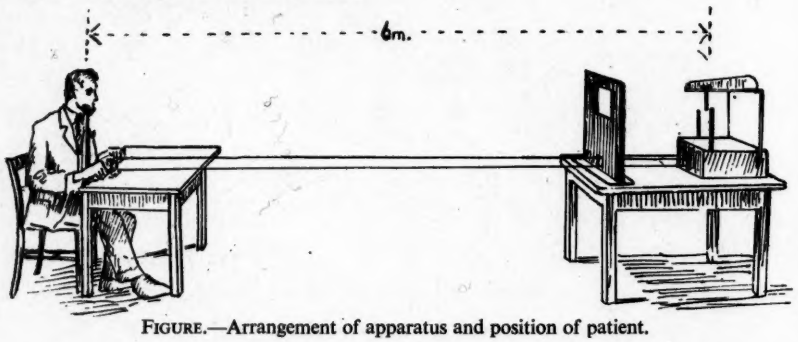

FIGURE.—Arrangement of apparatus and position of patient.

The subject's chin is placed on a chin-rest to prevent lateral movements of the head, so that motion parallax cannot furnish auxiliary clues to the relative positions of the three rods. The subject sees the rods as black silhouettes against a background of pearl glass, trans-illuminated at a level of 25 millilamberts. This level of background illumination should be kept constant throughout the examinations.

If the subject usually wears glasses for distant vision he should wear them during the test.

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The movable rod is controlled by the subject by means of a double string, one end of which is attached to the rod and the other to a wheel. This enables him to move the rod in three directions: up and down, from side to side, and to and fro.

At the beginning of a trial the administrator should place the movable rod at some distance nearer to or farther away from the subject than the stationary rods. This initial positioning is an essential factor in the procedure, because it may have a significant influence on errors of alignment and must be tested if accurate results are to be obtained (Wilner, Weymouth, and Hirsch, 1950).

The subject is required first to bring the movable rod forwards or backwards to align it laterally with the first stationary rod, then to move it sideways to align it vertically, and then to lower it so as to bring the ends of the two rods together to form a single vertical line. The starting position of the movable rod influences the final adjustments to apparent verticality, the apparent vertical tending to be closer to the first position in which the movable rod was set.

The subject is then required to carry out the same procedure with the second stationary rod.

The subject's powers of depth and height discrimination may be measured by his errors in aligning the rods, the stationary rods serving as points of reference. From a scale on the apparatus, the readings of successive attempts in the horizontal plane are noted in millimetres and the average reading is recorded. For vertical readings a narrow millimetre rule made of transparent plastic material has been devised. An average error greater than about 20 mm. on the vertical scale and about 30 mm. in the horizontal and lateral scales is indicative of subnormal powers of depth and height estimation. A single test-run comprising twelve adjustments of the movable rod should be allowed to accustom the subject to the apparatus. These initial readings are not a true indication of the subject's powers of depth and height discrimination. Reliability should not be sacrificed to speed or else the effort of testing may be useless.

From the twelve adjustments of the movable rod two independent scores may be computed: the average degree of accuracy obtained and the range of error about this average. The mean of the distribution expressed in millimetres gives the best estimate of the subject's powers. The standard deviation represents the degree of precision with which the subject approaches perfection.

In the Howard-Dolman test, the rods are viewed against an illuminated background through an aperture which conceals from view the ends of the rods. In the Zagora test, the lower end of the movable rod and the upper ends of the stationary rods are seen and therefore certain empirical clues to position are not eliminated.

The Zagora rod-test has been developed for determining the subject's fitness to undertake work in which a high degree of accuracy of binocular function and precision in depth and height perception and estimation, together with efficient ocular and manual co-operation are important.

The effects of artificial aniseikonic errors on this moving-rod test will be the subject of a future communication.

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Brit. J. Ophthal. (1957) **41**, 190.

CASE NOTE

TAY-SACHS'S DISEASE (AMAUROTIC FAMILY IDIOCY)*

BY

S. S. MANCHANDA, M. S. NIRANKARI, AND M. C. MAUDGAL

Medical College, Amritsar, India

The following cases are reported to draw attention to the occurrence of this rare condition in India.

Case Reports

Case 1, a male Hindu child, born at full term after a normal pregnancy and confinement history, was first seen at the age of 2 years in the Paediatric Department, V.J. Hospital. He had convulsions shortly after birth and was brought to hospital with a persistent respiratory illness. The mother reported that the baby had never been able to lift his head or sit up, recognize her, or take any interest in the world around him. He was the sixth in birth order. Three siblings were aged 7 and upwards and appeared normal, and two had died, one shortly after birth and the other at 8 months with convulsions before reaching the stage of being able to sit up.

The child was ill with bronchopneumonia, did not follow light, lift his head, or show any interest. He was able to move all four limbs. The deep jerks were brisk, and plantar reflexes flexor. Fundus examination showed clear media, normal discs, and at each macula a greyish-white area about 2 disc diameters in size with a typical "cherry-red" spot at the fovea. Laboratory tests were negative, but *x*-ray examination showed bilateral cystic disease of the lungs. The parents were not consanguineous.

Case 2, a male Hindu child aged 4, was brought to the same hospital on account of his inability to sit up or talk. He had been seen by doctors since the age of 8 months and pronounced mentally backward but the eyes had not been examined. He was the first and only child of unrelated parents and nothing abnormal was disclosed in the history of the family, the pregnancy, or the birth. Physically he thrived for the first few months. Examination showed that the child was poorly nourished with hypotonia and dystonic spasms of rigidity. Tendon jerks were present, and plantar reflexes flexor. There was no paralysis of any part of the body. He could follow light and recognize his mother but could not sit up or speak. Fundus examination showed a typical picture as in Case 1. All other pathological and radiological investigations were negative.

Discussion

The parents had no history of consanguinity, which is strictly forbidden by Hindu custom, and there was no Jewish element in these two Punjabi families. Both patients were males and the only suggestion of familial incidence was the death of a brother of Case 1 where a fundus examination might have shown the same macular picture.

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CORRESPONDENCE

OCULAR SYMPTOMS IN GLANDULAR FEVER

To the Editorial Committee of the BRITISH JOURNAL OF OPHTHALMOLOGY

SIRS—We are indebted to Dr. I. M. Librach for bringing to notice his two very interesting cases of glandular fever conjunctivitis (*Brit. J. Ophthal.*, 1956, **40**, 619), but his review of the literature is misleading. Dr. Librach asserts that there have been no reports of ocular manifestations of glandular fever in Great Britain since 1907 and that episcleritis has only been noted once before, and he implies that subconjunctival haemorrhages have not been previously described in this disease. He is surely mistaken about each of these three items.

The episcleritis, or circumscribed bulbar conjunctivitis, of glandular fever was vividly described by Chevallier and Bilski-Pasquier (1941). Subconjunctival haemorrhages were observed in glandular fever by Minot (1929 and 1936), and by Boger (1944), and they were recorded in Great Britain by Sears (1949). Tidy (1935), Stevenson and Brown (1943), and other authors have mentioned the not infrequent occurrence of conjunctivitis in glandular fever in Great Britian.

In 1955, in a discussion on lacrimal disease associated with infectious mononucleosis, I described an acute dacryo-adenitis in a Paul Bunnell-positive glandular fever which occurred in London. Some of the cases of dacryocystitis mentioned, but not described, were also true glandular fevers seen in London. The other cases in this report were seen in an epidemic of "Paul Bunnell-negative infectious mononucleosis" in Dunedin, New Zealand (Jones, Howie, and Wilson, 1952; Jones, 1954, 1955).

I should like to emphasize the distinction between the sporadic Paul Bunnell-positive cases seen in London and the negative epidemic cases seen in Dunedin (and similar cases seen in London), because it is likely that the latter group is due to a different aetiological agent or agents from the positive group. In other words, although similar, they are probably different diseases rather than differing reactions to the same disease, and it is therefore desirable (Leibowitz, 1953) to restrict the diagnosis of glandular fever to the Paul Bunnell-positive cases which give a characteristic absorption pattern of heterophil antibodies.

Yours faithfully,
BARRIE R. JONES.

INSTITUTE OF OPHTHALMOLOGY,
LONDON, W.C.1.
December 6, 1956.

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Brit. J. Ophthal. (1957) **41**, 192.

NOTES

Irish Ophthalmological Society

Annual Meeting, 1957

The Society of British Neurological Surgeons has accepted the invitation of the Irish Ophthalmological Society to a combined meeting of both societies to be held in Dublin on May 16, 17, and 18, 1957.

Dr. Frank Walsh (*Baltimore*) is delivering the Montgomery Lecture; his subject will be "Third Nerve Regeneration: Clinical Evaluation". A symposium on the blood supply of the chiasma and optic nerves has as openers Professor François (*Ghent*), Professor Erskine (*T.C.D.*), and Professor Brodie Hughes (*Birmingham*). Professor Chamlin (*New York*) is to speak on "Visual Function in *X*-Ray Treatment of Pituitary Tumours".

The Council of the Irish Ophthalmological Society extends a cordial welcome to all who wish to attend this combined meeting. The registration fee is half a guinea. Please notify the Secretary at the earliest possible date so that invitations, which are limited, may be issued.

L. B. Somerville-Large.

2, Fitzwilliam Place,
Dublin.

National Council to Combat Blindness

The National Council to Combat Blindness, in accordance with its programme concerned with the financing of research in ophthalmology and its related sciences, announces that applications for its 1957–58 Fight for Sight Grant-in-Aid and Fellowship Awards will be considered at the Eighth Annual Meeting of the organization's Scientific Advisory Committee to be held in the Spring of 1957.

The closing date for the receipt of completed applications for Grant-in-Aid and Fellowship Awards is *April 15, 1957.*

Applications for Summer Student Fellowships will be reviewed in advance of the meeting, and such applications should be filed with the office of the organization no later than *April 1, 1957.*

All applicants for fellowships are required to make their own arrangements for suitable facilities with accredited institutions.

Appropriate forms may be obtained from the Secretary, National Council to Combat Blindness, Inc., 30 Central Park South, New York 19, N.Y.

HONOURS

Her Majesty the Queen has sanctioned the appointment to the Order of St. John of Jerusalem, as Commanders, of A. J. Boase, O.B.E., F.R.C.S., and N. H. Ashton, *M.R.C.S.*

Air Vice-Marshal J. C. Neely, C.B.E., D.M., was appointed C.B. in the New Year *Honours.*

Sir Stewart Duke-Elder has been elected *Correspondant étranger de l'Académie royale de Médecine de Belgique.*

Mr. J. H. Dobree, F.R.C.S., has been appointed Ophthalmic Surgeon to St. Bartholomew's Hospital.